A STUDY ON ATAXIA

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CERTIFICATE

This is to certify that the dissertation titled **"A Study on Ataxia"** is a bonafide work done by **'Dr. A.K. SENTHIL KUMAARAN'.** It is a regular systematic study done under my guidance and supervision during the period of **18 months** (July 2004 to Jan 2006) and submitted for ensuring **'M.D Branch I General Medicine Examination**' September 2006 of Tamilnadu Dr. M.G.R. Medical University, Chennai.

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ABBREVATION

1. AT	:	ATAXIA TELENGIECTASIA
2. CAT	:	COMPUTRIZED AXIAL TOMOGRAPHY
3. CIDP	:	CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY
4. CP	:	CEREBELLO PONTINE
5. CVJ	:	CRANIO VERTEBRAL JUNCTION ANOMALY
6. DRPLA	:	DENTATO RUBRO PALLIDEO LUCEAN ATROPHY
7. EOCA	:	EARLY ONSET CEREBELLAR ATAXIA
8. FA	:	FREDRIECH'S ATAXIA
9. HCA	:	HEREDITARY CEREBELLAR ATAXIA
10. HMSN	:	HEREDITARY MOTOR SENSORY NEURONOPATHY
11. OPCA	:	OLIVO PONTO CEREBELLAR ATROPHY
12. SCA	:	SPINO CEREBELLAR ATAXIA
13.SCD	;	SUBACUTE COMBINED DEGENARATION

INTRODUCTION

"Taxia" means co-ordination in Greek. Ataxia therefore is defined as in-coordination or condition without order. They are mainly cerebellar, sensory and vertiginous ataxia.

They may be hereditary . congential or acquired. Hereditary ataxias are most often familial Sporadic cases do occur which may represent recessive inheritance in small kindreds or dominant inheritance with new mutations or variability of penetrance. It may also occur as a manifestation of a pleotropic gene.[14]

HISTORICAL ASPECTS

Friedreich was the first person to give its clinical description. comprising ataxia of the extremities, nystagmus and dysarthria appearing at puberty or earlier.

In 1891 Menzel described a family in which 4 members displayed ataxia of extremities followed at a later stage by dysarthria and choreiform movements and called this syndrome Olive Ponto Cerebellar Atrophy .

Ataxia telangiectasia was first described by Madam Louis Bar.

In 1972 Nakano et al described the proband Machado family residing in Massachussettes, which descended from a native of the island of Sao Miguel in the Portuguese Azores. The syndrome described included cerebellar signs, sensory loss and distal amyotrophy.

The hereditary ataxias may occur occasionally as one of the various phenotypic expressions of recognized inherited disorders. (For example, Type II Joseph diseasse Duveisin 1986) [34]. It may occur as dysgenesia in association with extraneural congenital and genetic defects. (For example a Syndrome of ectodermal dysplasia, short stature and hypogonadism, b DIDMOAD

–syndrome of Olivo Ponto Cerebellar Atrophy (Olivo Ponto Cerebellar Atrophy with diabetes Insipidus, diabetes Mellitus, Optic Atrophy and Deafness)[34] Hereditary ataxias may be dominant, recessive or sporadic.

Dominant Ataxias

Dominant ataxias are common. Intrafamilial variability is the rule rather than exception. The disease can begin at any age between 18 months to 80 years. But typically begins between adolescence and the forties. Ataxia involves the gait and extremities Ophthalmoplegia is often present and usually involves saccadic more than pursuit eye movements. Combination of pyramidal and extrapyramidal. dysfunction may be seen. Reflexes are typically increased early in the disease However, motor neuron degeneration occurs later in some patients, producing absence of tendon reflexes, atrophy and fasciculation. Dementia develops late in the course in some patients (Stupmf[68] 1985)

Recessive Ataxias

There are many distinctive forms of recessive ataxia. The most clearly defined variety is Friedreich ataxia. At least one third of recessively inherited ataxias do not fit in with the current clinically or biochemically defined diagnostic categories".

Sporadic Ataxias

Sporadic ataxias account for about one 3rd of the patients with chronic progressive ataxia[8].

Specific metabolic abnormalities have already been detected in some of these diseases and are assumed to cause many others as well.

Acquired Ataxias

Acquired Ataxias[67] can be produced by heterogeneous causes. Collectively they are the major group of ataxias. Important among these are vascular, neoplastic, infective, demyelinative and degenerative varieties.

Cerebrovascular diseases[67] producing acute and sub-acute infarctions, and hemorrhages may result in ataxic syndromes. Lesions of these types will result in cerebellar symptoms ipsilateral to the injured cerebellum and may be associated with impaired levels of consciousness due to increased intracranial pressure and ipsilateral pontine signs. Similarly, cerebllar tumors, demyelinating plaques of multiple sclerosis and abscess formation produceprogressive deficits ipsilateral to the cerebellar lesion. Two other important entities are paraneoplastics disease due to neoplasms outside the brain and causing bilateral cerebellar deficits by an antibody – mediated process and subacute cerbellar degeneration of the vermis with gait ataxia, due to chronic alcoholism.

Patients with AIDS[67] may develop an acute ataxia due to progressive multifocal leukoencephalopathy, which causes a rapid demyelination process, as a result of the JC virus. Progressive ataxia of gait and extremities may also caused by toxic or metabolic disorders, including hypothyroidism, hyponatremia. Vitamin B1 defeciency, vitamin B12 deficiency, toxic levels of phenytoin, lithium, bismuth, germanium, methyl mercury, and organic solvents and treatment with cyotoxic chemotherapeutic drugs. Rarely, congenital lesions, such as the Chiari type 1 malformation with cerebellar tonsillar compression of the brainstem, and congential dilatation of the fourth ventricle into a large cyst owing to impaired drainage of CSF (Dandy-Walker syndrome), present in adults as a progressive diffuse ataxic syndrome. Specific infectious diseases that can present with ataxia are meningovascular syphilis in patients recently infected diseases and tables dorsalis due to degeneration of the spinocerebellar pathway in chronically infected patients. Lyme's disease may cause ataxic symptoms.

Ataxias are an important problem of management with respect to diagnosis, prognosis and genetic counseling. Department of anatomy and Neurobiology, Washington.University school of medicine [59] in their study mechanism of ataxia (1997), they came to the conclusion that ataxia may be due to inability to co-ordinate the relative activity of multiple muscles and adjust movements at a given joint for the effects of other moving joints.

REVIEW OF ANATOMICAL AND PHYSIOLOGICAL ASPECTS

Many disorders present with ataxias but only three types of ataxias are of practical clinical importance sensory, cerebellar and vertegenous Sensory ataxia refers to dysfunction of posterior column, dorsal roots and peripheral nerves and a worsening of the ataxia once the sensory input is blocked. Sensory input from labyrinth are also important in maintaining postures. Cerebellar ataxia refers to incoordination due to cerebellar dysfunction. Because of the rich afferent and efferent connections from and to the rest of the C.N.S and of the unique anatomic arrangement of the organ, the cerebellum is well equipped to act as a servo system. Hence it is worthwile reviewing the functional anatomy of the cerebellum.

The cerebellum ("Small Brain [30(a)11]) is a dorsal portion of the metencephalon, lying in the posterior fossa of the cranium. The cerebellar surface has a characteristic patterning of parallel and curved furrows that separate the cortex into numerous laminae or folia. Each portion of the cerebellum has an anatomical name.

The narrow, central part of the cortex is called the vermis, the two larger, lateral masses are the right and left hemisphere. The several lobes and principal fissures of the cortex are identified as follow, in a retro-caudal direction:

1. Anterior lobe 2. Middle lobe

3. Posterior lobe 4. Flocculonodular lobe

The anterior lobe +pyramid+ uvula is known as the paleocerebellum, the middle and posterior lobes are the neocerebellum and the flocculonodular lobe+lingula is the archicerebellum. The anterior, middle, and posterior lobes are known as the corpus cerebelli. The hemisphere portions of the middle and posterior lobes are called, ansiform lobe, its relations are with the cerebro-pontine-cerebellar systems.

Afferent Fibres:

a. Climbing Fibres: *b.Mossy Fibres :*

Cells of the Cerebellar cortex:

a. Granule Cells b. Golgi type II neurons

c. Basket cells and Stellate cells d. Purkinje Cells

Cerebellar Nuclei:

a. Nuclues Fastigi b. Nuclei globosus c. embolifomis d. Dentate nucleus

CEREBELLER CONNECTIONS:

A. ***Inferior peduncle (restiform body) : Predominantly afferent***

B. ***Middle peduncle (brachium points):mainly afferent***

C. ***Superior peduncle (branchium conjunctivum): both afferent and efferent.***

Anatomy posterior Column[12a]

The large fibres, which subserve tactile and position sense and kinaesthesia project rostrally in posterior column on same side of the spinal cord and make their 1st synapse in the gracile or cuneate nuclei of lower medulla. The second order neuron decussates and ascend on the medial leminiscus located medially in the medulla and in the tegmentum of Thalamus. The third order neuron projects to the parietal sensory cortex.

Anatomy and Physiology of labyrinthine system[30b]

Receptors for 2 sensory modalities hearing and equilibrium are housed in the ear. The semi circular canals, the utricle, and the saccule of inner ear are concerned with equilibrium. Receptors in the semicircular canals detect rotational acceleration, receptors in the utricle detect linear

acceleration in the horizontal direction and receptors in the saccule detect acceleration in the vertical direction. The receptors are hair cell and they are 6 groups in each inner ear one in each '3' semi circular canal. one in urticle and one in saccule and one in cochlea (hearing).

Neural Pathyway[30b]

The cell bodies of the 19000 neuron supplying the cristae and macule on each side are located in thee vestibular ganglion. Each vestibular nerve terminates on the ipsilateral 4 part vestibular nucleus and in the floculonodular lobe of the cerebellum. Second order neurons pass down the spinal cord from the vestibular nuclei on the vestibulo spinal tract and ascend through the MLF to the motor nuclei of the cranial nerves concerned with the control of eye movement. The tracts that descend from the vestibular nuclei into the spinal cord are concerned primary with postural adjustments, and the ascending connections to cranial nerve nuclei are largely concerned with eye movements.

Pathogenesis of Unsteadiness

GAIT Imbalance [12b]

Imbalance results from disorders of the spinocerebellar of vestibular sensory impur, the integeration of these imputs in the brainstem or cerebellum, or the motor output to the spinal neurons that control axial and proximal muscles.

The position of the head in space is normally detected by the inner car. Excitatory input form the vestibular nerve and nuclei is to the fastigial nucleus deep in the midline of the cerebellum and via glutaminergic mossy fibres to overlying ipsilateral granule cells of the flocculonodular cerebellar cortex.

The position of the head relative to the limbs and trunk is detected by receptors that respond to joint position, joint movement and shortening or lengthening of muscle spindles in axial

and proximal limb muscles. This sensory input is transmitted both along the posterior column and medial lemniscal pathways to the cereberum and the spinocerebellar pathyways to the cerebellum. Visual input, by way of the tectum of the midbrain, is transmitted to the cerebellum through similar, excitatoty mossy fibres.

The midline cerebellar cortex and nuclei are of paramount importance in the integration of these inputs and in control of appropriated motor responses required to maintain normal balance.

The major cerebellar output for balance is from the fastigeal nuclei to the vestibular nuclei and reticular formation and, to a lesser extent, directly from the midline Purkinje cells to the vestibular nuclei, Vestibular nuclei and reticular formation project descending vestibulospinal and reticulospinal output via the ventromedial pathways to control the axial and proximal muscles of the limbs and trunk.

REVIEW OF LITERATURE

Several studies have been conducted in India[26,64,63] and abroad [8,49,65] on the ataxia.

HISTORICAL REVIEW

Nikolas Friedreich of Heodelberg in 1861 published as account of 9 cases of ataxia occurring in 3 families. It was then regarded as a hereditary or juvenile form of tabes or a form of multiple sclerosis. Again in 1876 Friedreich claimed it as an independent disorder and he called it as hereditary ataxia.

INCIDENCE

The first comprehensive clinical study of 32 cases of hereditary ataxia in India was done by Chuttani[26] et al from Amritsar between 1947 to 60. This study included 27 cases of Friedreich's ataxias. He emphasised that it was not an uncommon disease in India as was thought. He clearly described all the clinical features of the disease. He mentioned about a family of 8 siblings -3 with clinical evidence of Friedreich's Ataxia and 2" with forms fruste of Friendreich's Ataxia. In another family of 5 members, 3 were affected – 2 had features suggestive of Friedreich's Ataxia and one had Sanger Brown Marie Disease Jelly[63] in 1961 analysed 200 cases of spinocerebellar degeneration which included 9 cases of classical Friendreich's Ataxia, 6 of cerebellospinal group and 5 of cerebellar type. He included these cases under paraplegias because the patients primarily presented with difficulty in walking . A similar study of paraplegias was conducted by Chaudhary et al in 1968 and it showed a 9% incidence of spino cerebellar degeneration as compared to the 10% incidence in Jelly's series.

The prevalence of hereditary ataxia in Western countries varies between 7 and 17/100000 population (Werdelin[8], 1986).

Sahadevan[73] from Calicut studied 21 cases of during a period of 14 months (1963-64). His aim was to compare his findings with that of similar study conducted in a teaching hospital at London. He conducted that our patients had an earlier onset of cerebellar ataxia than that reported in the London series.

Wadia[64] and Swami studied 9 families with spinocerbellar ataxia and noticed slow eye movements in all of them.

Sarma and Vermani[61] (1972) reported 65 cases of spinocerbellar ataxia which included 2 cases of ataxia telangiectasia and one case of Ramsay Hunt syndrome.

CLASSIFICATIONS

In the course of time many attempts have been made to classify the hereditary ataxias clinically.

Holmles in 1910 classified hereditary ataxias into 3 groups

1. Predominant spinal form (Eg. Friedreich's Ataxia)
2. Cerebellospinal form (Eg. Sanger Brown Marie Disease)
3. Predominant Cerebellar form (Eg. Olive Ponto cerebellar Degeneration)

Later on in 1954, Green field modified this classification into 2 major varieties

1. Spinal form and 2. Cerebellar form

An indigeneous variety of spinocerebellar ataxia was described form India by Wadia and Swami[64] in 1971. They noted a particularly striking abnormally in their patients consisting of a severe loss of saccadic movement of the eyes. Skre and Loken[60] (1970) investigated a large number of Norwegian patients exhibiting hereditary ataxia, hereditary spastic paraplegia and Charcot-Marie-Tooth disease. Within the hereditary ataxia group they could distinguish between

Friedreich's ataxia in which the emphasis is on spinal symptoms, cerebellar ataxia with emphasis on cerebellar symptoms and spinocerebellar ataxia, a transient form between the other two. The starting point of his classification was genetic and clinical.

Harding (193)[49] studies about 274 cases of hereditary ataxias during 1966-80 and put forth a classification

1. Ataxias with known metabolic cause
2. Ataxias with no known metabolic cause.

Thus, as Refsum and Skree (1978) started there are as many classification as there are authors on the subject.Widely accepted classification at present are given below

Classification of Ataxia

1. Cerebeller ataxia 2. Sensory ataxia 3. Vertigenous ataxia

Classification of Cerebellar Ataxia (Hardings)[56]

1. Congenital Cerebellar Ataxia
   a. Ganule cell hypoplasia - Cerebellar ataxia, Mental Retardation
   b. Pontocerebellar hypoplasia - Cerebellar ataxia + MR + Spasticity
   c. Paine syndrome - Ataxia + MR + Spasticity – X- linked
   d. Gillespe syndrome - Ataxia + MR + Partial aniridia
   e. Joubert's syndrome - Ataxia + MR + Episode hyperpnoea

2. Congenital development anomaly

-CVJ anomly

-Basilar invagination

-Arnold Chiari malformation

3. Acquired Cerebellar Ataxia[71]

Trauma	
Tumours	
Vascular	Cerebellar haemorrhage
	Cerebellar infarct
	Vertebrobasilar TIAs
Infections	Tuberculoma
	Pyogenic / fungal abscess
	Acute viral cerebellitis
	Slow virus infections / prion diseases
	Falciparum malaria
	Enteric fever
Drugs	Anticonvulsants
	Antineoplastic agents
	Lithium
	Piperazine
Toxins	Heavy metals
	Solvents
	Organochlorine compounds
Nutritional / Alcohol	Vitamin B_1
(Deficiency of)	Vitamin E
	Vitamin B_{12}
Metabolic / Endocrine disorders	Hepatic encephalopathy
	Hypothyrodism
	Hypoglycaemia
Immune-mediated disorders	Multiple sclerosis

Post-infectious cerebellars

Miller Fischer syndrome

Paraneoplastic disorders

Hyperthermia/Hypoxia

4 Spinocerebellar ataxia – Genotypic Calssification[67]

SCA 1 to 22 – Autosomal Dominant

SCA 1 – Ataxia + Pyramidal + Extra pyramidal + Opthalmoparesis

SCA 2 - Ataxia + Pyramidal + Extra pyramidal + Slow saccades

SCA 3 - Ataxia + Pyramidal + Extra pyramidal + Amyotrophy

SCA 4 - Ataxia + Pyramidal + Sensory neuropathy

SCA 5 - Ataxia + Dysarthria

SCA 6 - Ataxia + Dysarthria + Nystagmus

SCA 7 - Ataxia + Retinal Degeneration

SCA 8 - Ataxia + Dysarthria + Nystagmus + Leg spasticity + Reduced vibratory sensation

SCA 10 - Ataxia + Dysarthria + Nystagmus + Seizures + Polyneuropathy

SCA 11 - Ataxia + Dysarthria + Vertical Nystagmus + Hyper reflexia

SCA 12 – Ataxia + Tremor + Decreased movement + Dementia +Dysautonomia

SCA 13 - Mutation unknown

SCA 14 - Mutation unknown

SCA 15 - Ataxia + Dysarthria

SCA 16 - Ataxia + Dysarthria + Head tremor + Horizontal nystagmus

SCA 17 - Ataxia + Dementia + Seizures + Parkinsonism

SCA 18 - Ataxia + Sensory neuropathy

SCA 19 - Ataxia + Tremor + Cognitive impairment + Myoclonus

SCA 20 – Assigned not yet published

SCA 21 - Ataxia + Tremor + Cognitive impairment + Rigidity

SCA 22 - Assigned not yet published

DRPLA – Ataxia + Dementia + Dystonia + Myclonus + Choreoathetosis+ Seizures

Episodic Ataxia (AD)

Type -I Ataxia for minutes Type –II Ataxia for days

Freidreich's Ataxia (AR)

5. Ataxia Due to Mitochondrial Disorders[67]

1. MERRF syndrome – AR

Myoclonic + Epilepsy + Ragged red fiber myopathy + Ataxia

2. MELA'S syndrome – AR

Mitochondrial encephalopathy + Lactic Acidosis + Stroke + Ataxia

3. KEARN'S syndrome (AR)

-Opthalmoplegia + Hear Block + RP

4. LEIGH'S syndrome

- Hyptonia + Obtundation + Respiratoty failure

5. ATAXIA TELENGIECTASIA

- Telengiectasia + ataxia + Respiratory infection

6 Hereditary Ataxia due to metabolic cause[71]

Intermittent Ataxias

With hyperammonaemia

Urea cycle defects

Maple syrup urine disease

Hartnup's disease

Isovaleric aciduria

Disorder of pyruvate / lactate metabolism

Pyruvate dehydrogenase deficiency

Pyruvate carboxylase deficiency

Multipile biotin dependent carboxylase deficiency

Mitochondrial disorders

Progressive Ataxias

Ataxia occurs as a major feature

Ataxia with isolated vitamin E deficiency (AVED)

Abetalipoproteinaemia

Cerebrotendinous xanthomatosis

Refsum’s disease

Hexosaminidase deficiency

Ataxia occurs as a minor feature

Wilson’s disease

Neuronal ceroid lipofuchsinosis

Leucodystrophies

Sialidosis

ETIOLOGY OF CEREBELLAR ATAXIA[67]

Symmetrical and Progressive sign			**Focal and ipsilateral Cerebellar signs**		
Acute	Subacute	Chronic	Acute	Subacute	Chronic
Alcohol, lithium Diphenylhydantoin barbiturates	Intoxication: Mercury, Gasoline, Chemotherapeutic drugs	Paraneo plastic syndrome	Vascular, Cerebellar infarction, hemorrhage, or subdural hematoms	Neoplastic cerebellar glioma or metastatic tumor	Stable gliosis secondary to vascular lesion or demycoinating plaque.
Acute Viral Cerebellitis	Alcoholic-nutritional	Hypothyroidism	Infections Cerebellar abscess	Demyelinating Multiple sclerosis Aids related PML.	Congenitial lesion Dandy-Walker of Arnold-Chiarimalfornations.
Postinfection Syndrome	Lyme's disease	Inherited Diseases			

CLINICAL FEATURES

The subject of Olivo-Ponto-Cerebellar atrophy has been extensively reviewed by Konigsmark and Weiner [41] (1970) has been classified into 5 subgroups.

S.No	Types	Clinical Features
1	Type I	Autosomal dominant – Menzel type
2	Type II	Autosomal recessive – Fickler Winkler type
3	Type III	Autosomal dominant – Olive Ponto Cerebellar Atrophy with retinal degeneration
4	Type IV	Autosomal dominant – Schut kindred
5	Type V	Autosomal dominant – Ophthalmoplegia Dementia and extrapyramidal manifestation

The essential features of Olivo-Ponto-Cerebellar Atrophy are cerebellar and extrapyramidal dysfunction-Duvoisin,[34] 1986. The classical eye signs in Olivo-Ponto-Cerebellar Atrophy are hypometric Saccades, impairment of upward gaze, loss of optokinetic nystagmus. (Duvoisin, 1986). Supranuclear ophthalmoplegia has also been reported in dominant ataxia. Slow saccades were characteristically described in an indigenous variety of ataxia by Wadia Swami (1971). Garcin and Man in 1958 described the characteristic slow eye movement seen in cerebellar and spino cerebellar degeneration.

Rendent et al[52] (1983) reported a case of Menzel's ataxia with slow eye movements myoclonus, facial dystonia and signs of spinal cord and peripheral nerve involvement Neuropathological examination revealed Olive Ponto Cerebellar Atrophy associated with degenerative changes of spinal cord characteristic of Menzel's ataxia. Slow eye movement in the case could be specific for one type of Olivo-ponto-Cerebellar Atrophy. The horizontal voluntary saccadic eye movements were regularly slow and more severely impaired than vertical movements. Horizontal pursuit movements were less affected than saccades.

One of the distinct variety of OPCA is familial, essentially autosomal dominant cerebellar ataxia associated with slow saccadic eye movements. It is now commonly known as 'Wadia type' of OPCA. Wadia so far reported 60 members (38 males and 22 females) and 23 families from Maharashtra belonging to Hindu, Muslim and Christian religions. The south Indian family of Kini and Venugopal also had similar oculomotor disorder with cerebellar ataxia and similar autosomal dominant pattern of inheritance, although oculometic examination s and autopsy were not done in these cases.

Neuroimaging[42] with CT and MRI shows ballooning of fourth ventricle, because of excavation of its floor together with atrophy of brachia pontis and conjuncitva give a characteristic 'molar tooth' appearance with its root projecting posteriorly (Wadia, Biswas, S.Singh – 1997)

In Wadia's view "this is the most common variety of ataxia in India, not excluding Friedreich's ataxia". However, all reported cases so far from India are restricted to Maharashtra state except the family of Kini and Venugopal from South India.

A comparative study of familial and sporadic Olive Ponto Cerebellar Atrophy was done by Bercanio[72] (1982). He gathered 54 cases of familial Olivo Ponto cerebellar Atrophy from literature. He found that the disease began earlier in familial Olivo Ponto Cerebellar Atrophy and lasted longer. He stressed that the differences in the percentage of clinical manifestation and associated lesions were also significant with regard to the greater frequency of abnormal movements, ophthalmoplegia, spinal symptoms and lesions in the dentate nucleus and spinal cord in familial cases.

Bercanio[72] (1982) described urinary incontinence in advanced stage of Olive Ponto Cerebeller Atropohy. He found that among 117 cases, 51 had urinary incontinence of whom 28

had dementia and 27 had posterior column degeneration. Urinary retention was extremely uncommon, but double incontinence was not rare.

Stumpf[68] (1985) has reported the occurance of neurogenic – Spastic and atonic bladders in several of their patients with Friedreich's Ataxia.

Dysphagia had been described as an important symptom in the intermediate and advanced stages of the Olivo Ponto Cerebellar Atrophy except from occasional early occurrence (Bercanio 1982).

These changes according to Duvoisin[34] consistent with lesions of locus ceruleus and pontine tegmentum. Sleep abnormalities well documented in OPCA by Duvoisin – 1986. These disorders reflect neuronal degeneration in brainstem regions where hypnogenic and respiratory control mechanisms are situated.

SCA2 Symptoms and sign

Another clinical phenotype, SCA2, has been described in Cubans. These Patients Probably are descendants of a common ancestor, and the population may be the largest homogeneous group of ataxic patients yet described. The age of onset ranges from 2 to 65 and there is considerable clinical variability within various families.

Machado-Joseph Disease / SCA 3:

Machado- Joseph – Azorean Disease:

After the first description by Nakano et al (1972) the clinical and pathological features of Machado-Joseph-Azorean disease have been studied by several workers[51&50]. In 1972 itself Woods and Schaumbur[66] described a family of Portuguese descent having nigro spino dentatal

degeneration with nuclear opthalmoplegia, characterized by gait ataxia, ophthalmoplegia, spasticity or rigidity in the limbs, nystagmus and limb ataxia.

Rosenbergy[50] (1976) reported a Portuguese family in which clinical signs were different. Spasticity, lurching gait, spastic dysarthira, loss of fast saccade eye movement, opthalmoparesis for upward gaze, facio-lingual fasciculations and dystonia without signs of cerebellar dysfunction were found. The neuropathologic features included degenerative changes of substantia nigra, Clarke's column, Anterior Horn cell and involvement to a lesser degree of Pontine nuclei and brainstem cranial nerve nuclei, sparing the inferior olives. Decrease in Homo Vanilic Acid levels in Cerebro Spinal fluid has been reported in Joseph's disease and it is due to marked nigral degeneration seen in this disease (Sakai[51] et al, 1983). Roseenberg et al (1976) have studied protein patterns in fibro blast and brain in patients with joseph disease and have reported increase in J or L. proteins in frontal cortex. cerebellar cortex and putamen (1981).

Barucha et al[15] (1986) described a syndrome very similar to Joseph's disease from an Indian family in which 3 generations had varying combinations of ophthalmo paresis cerebellar ataxia, puramidal signs, amyotrophy and intentional facial fasciculation like movements, as the salient clinical features.

MID has been found in families from Portugal, Australia, Brazil, Canada, China, England, France, India, Italy, Japan, Spain, Taiwan and the United Stares.

Sakai et al[51] in 1983 reported a family showing 2 types of neurological abnormality – one was dominated by cerebellar and pyramidal signs, loss of fast saccades, horizontal and vertical nystagmus, opthalmoplogia with negative oculocephalic rerlex and facio-lingual faciculations, all characteristic of Machado-Joseph disease type II. The second patient had cerebellar signs peripheral sensory loss, loss of tendon reflexes – features compatible with type III Joseph disease.

Autopsy of these cases showed marked degeneration of the substantia nigra, anterior horn cell, Clarke's column and dentate nucleus with involvement of pons and cranial nerves or their nuclei.

Abnormalities of saccadic eye movement were also described by Healton et al (1979) in 4 patients of a black family in 1979.

SCA 4

One family with progressive ataxia, pyramidal tract deficits, normal eye movements, and prominent sensory axonal neuropathy is described in which the trait has autosomal dominant transmission and is mapped to chromosome 16q24-ter.

SCA 5

Finally, another family is reported (which has two major branches, both descended from the paternal grandparents of President Abraham Lincoln) in which dominantly inherited spinocerebellar ataxia (SCA5) is mapped to chromosome 11.

SCA6

Yabe-1, Sasaki et al[47]1998 (Department of Neurology, University school of medicine – Hokkaido – Japan) analysed the initial symptoms and the mode of progression in this disorder on 25 genetically verified patients. The initial symptoms were recurrent episodes of transient vertigo (72%) or unsteady gait (28%), gaze evoked nystagmus (92%) transient positional nystagmus (82%) and periodic alternating nystagmus (4%) in addition to cerebellar ataxia. These fluctuating symptoms at the initial stage of the illness were clearly different from those to other SCA. They also stressed the point the clinical similarly between SCA 6 and Episode ataxia type II and suggested that there might be a common mechanism.

Genomic screening for CAG repeats in other families with autosomal dominant ataxia have yielded another locus.

SCA 7

Retinopathy associated with familial cerebellar atrophy was published by Carpenter[21] and Schumacher (1996). Optic atrophy and opthalmopelgia were observed by Weiner and Kogniamark (1970) in 27 affected individuals of 5 generations in a family. Only 14 of these patients were examined and the remaining 13 were presumed to have ocular involvement by reliable history. 14 had pigmentary changes, 10 had optic atrophy and 5 had ophthalmoplegia.

SCA – 8 to 25

Studies are going on new types of spinocerebellar ataxias. Their current number is large and includes SCA 1 through SCA 25[75]. SCA 12 is one of the recently identified SCA's , first described by Holmes, O'Hearn and colleagues in 1999. In a phenotype genotype study conducted at Ranchi on 54 families with ADCA, 12 famalies with SCA 12 mutation was identified[76]. Mutation produced expanded CAG repeats, smallest reported was 51 CAG repeats (Srivastava et al 2004)[77]

DRPLA

Sirichai and Walters in 1984 reported an autosomal Dominant syndrome of progressive sensori – neural deafness, myoclonus and cerebellar ataxia in identical twins of a family and 3 other members of the family had hearing loss and 2 had seizure disorder. Autosomal dominant late onset cerebellar ataxia with myoclonus, peripheral neuropathy and sensori-neural deafness was also published by Baraister et al (1984).

Skre and loken (1970)[60] also published a report of a family with 3 members having ataxia. One member had epilepsy along with ataxia the other had dementia and yet another had Schizophernia. On following them up one brother of the affected patient also later on developed myoclonic epilepsy and progressive dementia along with ataxia.

Bird and Shaw (1984)[17] also reported the association of progressive myoclonus and epilepsy with dentatorubral degeneration. The pathological findings reported by the autors varied considerably but the involvement of dentate nucleus was almost always invariable, with severe neuronal loss and gliosis. (Baraitser et al, 1984).

Van Bagaert ans Martin (1984)[18] studied a families with hereditary ataxias and came across several findings like deafness, optic atrophy, albinism, mental disturbances, endocrine abnormalities and pigmentary disorders apart from ataxia. They commented that in Hereditary Ataxia, disturbances of the vestibular apparatus were more common in as many as 2/3rd of the patients with Hereditary ataxia. Temporal bone pathology was reported in 2 cases of Friedreich's ataxia with vesibulo-cochlear-disorders by Spoendlin (1974). He observed selective degeneration of the primary neurons with predominant damage to the cochlear nerve and greatest preservation of the neurons of the macular branches of the vestibular nerve.

Ramsay Hunt Syndrome

Now Ramsay Hunt syndrome is considered to be a mitochondrial cytopathy (Petty et al, 1986[46] and Dimauro et al, 1985[33]) May and White[62] (1968) reported the occurrance of cerebellar ataxia. deafness and myoclonic jerks in several members of a single family. It was thought to represent a distinct, genetically determined disease transmitted by an autosomal dominant gene.

Episode Ataxia Types 1 And 2

These are two rare dominantly inherited disorders that have been mapped to chromosomes 12p (a potassium channel gene) for type I and to 19p for type 2.

Mitochondrial Ataxias

Spinocerebellar syndromes have been identified with mutations in mitochondrial DNA (mt DNA). Thirty pathogenic mt DNA point mutations and over 60 different types of mt DNA deletions are known, and several of these mutations cause or are associated with ataxia.

Ataxia Telangiectasia

The clinical syndrome of ataxia telangiectasia has attracted attention of various research workers. In 1963 Cutman[31] reported 2 cases of ataxia tenlangiectasia. Another study about this was done by Mckusick and Cross[45] in 1966. In a family 2 members had full blown disease and another member had only Swiss type of agammalgobulinaemia. Cutaneous manifestation of this disease were studied in detail by Reed[44] in 1966. In 1965 Karpate and Isen reported 6 patients of 4 unrelated families 6 were alive and 2 dead. All of them had dysgammaglobulinaemia.

Millar[43] (1969) reported 2 cases of ataxia telangiectasia among 28 cases of cerebellar disorders of childhood. Hong and Amman (1970) demonstrated antinuclear antibody in this disease and suggested that it was an auto-immune disease affecting multiple organs.

Amman in 1969 stressed that patients of ataxia telangiectasia wre more prone to respiratory infection. They had in addition to lg G deficiency, lg E deficiency also. In 1969 Verma published a report of Ataxia telangiectasia, emphasising the clinical features of the disease and the dysgammaglobulinaemia associated with it.

Pathologically the Purkinje and granular cells of the cerebellum are selectively involved in ataxia telangiectasia (Walton, 1986).

Martinez et al (1977) demonstrated abnormalities of sensory and mixed evoked potentials with other evidence of peripheral nerve involvement in Ataxia telangiectasia. Teplitz (1978) stressed that patients with ataxia telangiectasia showed marked deficiency of DNA repair and that the condition was characterized by spontaneous chromosomal instability.

Friedreich's Ataxia

Nanning[40] in 1950 studied 5 patients with Friedreich's ataxia – 3 patients showed abnormal rhythym and extrasytoles. One had cardiac failure and one showed T wave abnoramalities in the electro cardio gram.

The 1962 Boyer et al analysed electro-cardio graphic abnormalities in 38 cases of Friedreich Ataxia. Among these, 12 had abnormal electro-cardio graphic findings such as T inversion, low voltage complexes, multiple ectopics and diphasic T.

9 cases of Friedreich Ataxia with Cardiac manifestation in children were reported by Adams and Anderson (1955). Cardiomyopathy, generally of the hypertrophic type was described as a cardinal feature of Friedreich's Ataxia by various other workers also (Cote[28] et. Al., 1976).

ACQUIRED ATAXIA

Cerebellar Ataxia

Among the various causes producing cerebellar ataxia, cerebro vascular accident is the most common. It can be due to thrombosis or embolism.

CVA Affecting Cerebellum and Brain Stem

The northern Manhatton[70] stroke study done by Gan R, Sacco RL, Kagman et al in 1997 categorised lacunar syndrome as pure motor hemiparesis (PMH), pure sensory syndrome (PSS) sensory motor syndrome (SMS), ataxic- hemisphere (AH) and other lacunar syndromes and found out that PMH was the commonest lacunar syndrome accounting for 45%, SMS 20%, AH 18% and PSS 7%.

Among the 196 patients with lacunar syndromes they studied, atherosclerosis accounted for 17 (19%), Cardioembolism 10(15%), Cryptogenic 17(9%) and other usnusual causes 4(2%)

Atherosclerosis has a predilection for the origin and the distal segments of the verteberal arteries, the proximal basilar artery, and the origin of the major and minor branches of the vertebral, basilar, and posterior cerebral arteries. Predictably atheromatous disease at each site produces its own clinical syndromes. Caplan LR[69], Pressum MS and Mohr JP (1992) stressed the point atherosclerosis is the commonest etiology of vertebro basilar occlusive disease and embolic manifestations are rare but can occur.

Atheroma in the fourth segment of the vertebral artery can occur proximal or distal to the origin of the posterior inferior cerebellar artery, as well as at the junction with the other vertebral artery that forms the basilar artery. When it is proximal to the origin of the posterior inferior cerebellar artery, a critical narrowing can threaten the lateral medulla and posteroinferior surface of the cerebellum[38].

Atheromatous occlusion of the penetrationg branches of the thalamic and thalamogeniculate arteries)Castaiqe P, Lhermitte F, Buge et al - 1981[37]) produces less extensive thalamic and thalamocapsular lacunar syndromes. The thalamic syndrome of Dejerine and Roussy is the best known. Its main feature is contralateral hemisensory loss of both superficial sensation

(pain and temperature) and deep sensation (touch and proprioception). Occasionally, it may affect only pain and temperature or vibration and joint position sense. After a few week or months, an agonizing, searing or burning pain may develop in the affected areas. Weisberg LA (1986)[36] re-analysed this syndrome by clinical and C.T. Correlation study of thalamic hemorrhage.

Vertebral and Posterior Inferior Cerebellar Arteries:

TIA's resulting from vertebral artery insufficiency cause dizziness or vertigo, numbness of the ipsilateral face and contralateral limbs, diplopia, hoarsenss, dysarthria, and dysphagia. Hemiparesis is rare.

Superior Cerebellar Artery[73]

Occlusion of this artery results in severe ipsilateral cerebellar ataxia, nausea and vomiting dysarthria, and contralateral loss of pain and temperature sensation over the extremities, body and face.

Anterior Inferior Cerebellar Artery:[73]

Occlusion produces variable degrees of infarction because the size of the artery and the territory it supplies vary inversely with those of the posterior inferior cerebellar artery. The principal symptoms include ipsilateral deafness, facial weakness, true vertigo.

LacunarDisease

Amarenco O and Hauw JJ's [27]study (1990) of 20 cases of cerebellar infarction in the territory of anterior inferior cerebellar artery and posterior inferior cerebellar artery.

On side opposite to lesion:

Impaired pain and thermal sense over half the body, sometimes face (Spinothalmic tract).

Total unilateral medullary syndrome (occlusion of vertebral artery). Combination of median and lateral syndromes.

Lateral pontomedullary syndrome (occlusion of vertebral artery). Combination of lateral medullary and lateral inferior pontine syndrome. The very rare case of lateral inferior pontine syndrome and lateral medullary syndrome due to vertebral artery dissection was reported in 1977 by Hashimot Y et al[29] from Department of Neurology, Kumamato city hospital.

Basilar artery syndrome (the syndrome of the lone vertebral artery is equivalent) A combination of the various brainstem syndromes plus those arising in the posterior cerebellar artery distribution.

a. Bilateral long tract signs (Sensory and motor; cerebellar and peripheral cranial nerve abnormalities);
b. Bilateral long tact; cerebellar and peripheral cranial nerves.
c. Paralysis or weakness of all extremities, plus all bulbar musculature.
   Corticobulbar and corticospinal tracts bilaterally.

Ataxic hemiparesis from vascular lesions of corona radiata also reported by sage JI et al in 1983. Masson C, Berhtelot et al (1997)[55] reported case of ataxic crural hemiparesis caused by posterior spinal artery infarction. Infarction in the territory of PSA is uncommon, giving rise to softening of cord of variable extent.

Cerebellar Hemorrhage

This usually develops over a period of several hours and loss of consciousness at the onset is usual. Repeated vomiting is a prominent feature, along with occipital headache vertigo, and inability to sit, stand, or walk. Often these are the only abnormalities making it imperative to have the patient stand and walk. Otherwise the examination may appear falsely normal. In the early phase of the illness, other clinical signs of cerebellar disease may be minimal or lacking, only a minority of cases show nystagmus or cerebellar ataxia of the limbs, although these sign must always be sought. A mild ipsilateral facial weakness and a diminished corneal reflex are common. Dysarthria and dyphagia may be prominent but certainly may be absent. Contralateral hemiplegia and facial weakness do not occur unless there is horizontal displacement of the medulla against the clivus. There is often paresis of conjugate lateral gaze to the side of the hemorrhage, forced deviation of the eyes to the opposite side, or an ipsilateral sixth nerve weakness. Vertical eye movements are retained other ocular signs include blepharospasm, involuntary closure of one eye skew deviation. “Ocular bobbing,” and small but often unequal pupils that continue to react until very late in the illness.

INFECTIONS

Cerebellar Abscess

Pathogenesis:

With the exception of a small proportion of cases (about 10 percent) in which infection may be introduced from the outside (compound fractures of the skull, intracranial operation, bullet wounds), brain abscess is always secondary to focus of suppuration elsewhere in the body. Approximately 40 percent of all brain abscesses are secondary to disease of the paranasal sinuses, middle ear, and mastoid cells.

Otogenic and rhinogenic abscesses reach the nervous system in one of two ways. One is by direct extension, in which the bone of the middle ear or nasal sinuses becomes the seat of an osteomyelitis,.

It is estimated that about 5 percent of cases of congenital heart disease are complicated by brain abscess (Cohen[25], Newton, 1966). In children, more than 60 percent of disease (Malson), the tetralogy of Fallot is by far the most common anomaly associated with brain abscess but the latter may occur with any right – to left shunt. Nearly half of the reported cases of pulmonary arteriovenous fistulas also have Osler-Rendu-Weber telangiectasia and neurologic symptoms. Without telangiectasia. Only 18 percent have neurologic symptoms of these 5 percent prove to have brain abscesses.

A multicentre prospective study about bacteriology of abscess of CNS by DeLovois et al[24] came to conclusion that streptococci are the commonest organism responsible for cerebellar abscess.

Headache is the most frequent initial symptom of intracranial abscess. Other presenting symptoms, roughly in order of their frequency, are drowsiness and confusion.

Acute Cerebellitis (Acute Ataxia):

Ataxia is a component of both infections and postinfectious encephalitis, but a special comment should be made concerning the isolated acute ataxia due to meningocerebellitis. It was described by Westphal in 1872, following smallpox and typhoid fever in adults, but Batten is credited with defining the more common ataxic illiness that occurs after childhood infections of measles, pertusis and scarlet fever. Currently acute ataxia is most often associated with chicken pox (one quarter of 73 consecutive cases reported by Connolly er al), but it can occur after (or together with or just before) any of the exanthems, as well as with enterovirus (mainly

Coxsakie)EBV, mycoplasma, cytomegalovirus, Q fever, vaccine, a number of vaccinations, rarely with HSV, and also after nondescript respiratory infections (Weisis and Guuberman). In adults the most common preceeding organisms are probably EBV and mycoplasma. The syndrome appears relatively abruptly, over a day or so, and consists of limb and gait ataxia and less consistently, dysarthia and nystagnus. Often there are additional minor signs such as increased limb tone, Babinski's sign or confusion. The spinal fluid shows a mild pleocytosis and the protein is elevated but some cases have a normal CSF. MRI show no abnormality in the majority of cases. Most patients make a slow recovery, but permanent residua are known to occur. Brownell[23] and oppenheimer reported an ataxic form of creutzfeldt – jakob disease in 1965.

Malaria with Ataxia

The study of residual neurologic sequelae after childhood malaria done by Vanhensbroek et al,[22] in Royal Victoria hospital, UK (1997) came to some important conclusion about cerebral malaria. The prospective study in 624 patients, admitted with cerebral malaria in two hospital. By one month the proportion had decreased to 86% and at 6 months only 4.4% of the survivors sequelae. The most common form of neurologic sequelae were paresis, and ataxia often found in combination with other neurologic abnormalities.

Neuroborreliosis[19]

Zafkowska et al (1998) reported a case of 47 yrs old female with progressive hearing loss, tinnitus, ataxia and paraparesis following borrelial infection. Her CSF showed mononuclear pleocytosis, protein concentration over 600 mg% and antibodies against borrelia burgodorgeri in 1gG and lgM class.

Brain Stem Tumor

Astrocytomas of the brain stem are relatively slow growing tumours that infiltrate tracts and nuclei. They produce a variable clinical picture, depending on their location in the medulla, pons or midbrain.

(A careful imaging and clinical study of 87 patients by Barkovich[20] and coworkers (1993) has emphasized the importance of distinguishing between diffusely infiltrating and focal nodular tumors. The diffusely infiltrating tumours, usually showing an asymmetrical enlargement of the pons, have a poorer prognosis than the focal or nodular tumors). With the conclusion of study, they made a classification of Brainstem glioma based on MRI.

Acoustic Neuroma (Vestibular Schwannoma)

This tumor was first described as a percentage entity by Sandifort in 1777, first diagnosed clinically by Openheim in 1890, and first recognized as a surgically treatable disease around the turn of the century. Cushing's monograph[16] (1917) was a milestone, and the papers of House and Hitselberger and of Ojemann and colleagues provide excellent description of the modern diagnostic tests and surgical treatment as well as comprehensive bibliographies.

A detailed study about clinical feature of acoustic neuroma done by Harner[13] S.G. They noted, as the eigth nerve schwannoma grows, it extends into the posterior fossa to occupy the angle between the cerebellum and pons (cerebellopontine angle). In this lateral position it is so situated as to compress the seventh, fifth, and less often the ninth and tenth cranial nerves, which are implicated in various combinations. Later it displaces and compresses the pons and lateral medulla and obstructs the CSF circulation very rarely, it is a source of subarachnoid haemorrhage. (Harner S.G.,Laws ER – Mayo Clinic Proc 1983).

Cushing H.(1917) in his monograph stressed that there are so many space occupying lesions other than acoustic neuroma to produce C.P angle syndrome like, meningioma, cholesteatoma, secondaries, tuberculoma and even syphilitic gumma.

Degenerative Disorders

Neurodegenerative disorders, (Joseph.B, Martin et al[11] 1999) which are chronic and progressive, are characterized by selective and symmetric loss of neurons in motor, sensory, or congnitive systems. Delineation of the patterns of cell loss and the identification of disease-specific cellular markers have aided in nosologic classification.

Important degenerative disease affecting cerebellum are hereditary cerebellar ataxia, alcoholic cerebellar degeneration and paraneoplastic degeneration of cerebellum.

Alcoholic Cerebellar Degeneration

This terms refers to a common and uniform type of cerebellar degeneration in alcoholics. This disorder is about twice as frequent as Wernicke disease, but unlike the later, it is considerably more frequent in men than in women. According to Trovik et al (1982), it is characterized clinically by a wide-based stance and gait. Varying degree of instability of the trunk, and ataxia of the legs, the arm being affected to a lesser extent and often not at all. Nystagmus and dysarthria are infrequent signs. (Torvik A. Lindboe- 1982)[58]

Behse F and Buchthal F (1977)[10], in their biopsy study about alcoholic cerebellar degeneration, demonstrated the pathologic changes which consists of a degeneration of all the neurocellular elements of the cerebellar cortex, but particularly of the Punkinje cells, and are restricted to the anterior and superior aspects of the vermis and in advance cases, to the anterior parts of the anterior lobe. The cerebellar atrophy is readily visualized by CT scanning.

There are two particular forms of this syndrome. In the chronic fixed form, the clinical abnormalities are limited to an instability of station and gait. A second type is acute and transient in nature: except for their reversibility, the cerebellar symptoms are identical to the ones that characterize that chronic, fixed form of the disease.

Victor M and Mancall El (1959) mentioned a restricted form of cerebellar degeneration in alcoholic partients in their study. The cerebellar ataxia of Wernicke disease and that referred to as alcoholic cerebellar degeneration represent the same disease process. (Adams).

Paraneoplastic Cerebellar Degeneration

. In revieweing this subject in 1970 it was found that only 41 were pathologically verified cases (Adams), and in a subsequent review (Henson and Urich 1982)[9] only a few more case were added. The actual incidence is obviously higher than these figures indicate. At the Cleveland Metropolitan General Hospital, in a series of 1700 consequtive autopsies in adults, there were five instances of cerebellar degeneration associated with neoplasm. In the experience of Henson and Urich, about half of all the patients with nonfamilial, late onset cortical cerebellar degeneration proved sooner of later to be harboring. In recent years, large series of cases been reported from mayo clinic and the Memorial Sloan-Kettering Cancer Center (Hammock et ak, Anderson et al).

In approximately one-third (33%) of the cases, the under–lying neoplasm has been in the lung (most often a small-cell carcinoma) this figure reflects the high incidence of this tumor.

Characteristically the cerebellar symptoms have an insidious onset and steady progression over a period of weeks to months, in more than half the cases, the cerebellar signs are recognized before those of the associated neoplasm.

Anderson NE., Rosenblum and Posner[6] have done a study about clinical and immunological correalation in paraneoplastic cerebellar degeneration in 1988

CV Junction anomaly:

Def. craniovertebral anomalies [53] are developmental defects involving the bony and or neural structure at the occipito cervical transition zone. This is one cause of cerebellar ataxic syndrome; which is due to a congenital anomaly that later on leads to neurological manifestations. This can be due to bony pathology or soft tissue pathology Bony pathologies are platybasia, basilar invagination, Klipped fail syndrome, atlantoaxial dislocations and occpitalistation of atlas. Soft tissue anomalies are Arnold chiarimalformation. Dandy-Walker syndrome, occipitocervical meningomyelocele and cysts in the posterior cranial fossa. Combined anomaly can also be present.

Common clinical features are short neck (Height Neck Radio, > 13.86) low hair line, facial asymmetry and restricted neck movements.

Secondary changes such as repeated trauma pressure effects and vascular occlusions aggravate the neurological disability and lead to progression.

This syndrome can be proved radiologically by taking X-ray skull AP, lateral view and open mouth view, X-ray neck in flexion, neutral position and in extension. Various radiological lines used are MC Gregor's line., Bimastoid line, digastric line and chamber lane's line.

Sensory Ataxia

Preservation of upright position depends upon labyrinthine, cerebellar and visual postural reflexes and upon those reflexes whose afferent pathway is from the proprioceptors of lower limb. As long as the eyes are open even if the conduction of proprioceptive stimulus is grossly impaired, the patient can maintain balance, but once eyes are closed the will sway or fall (Romberg's sign).

Sensory ataxia is differential from cerebellar and labyrinthine ataxia in that severe instability and tendency to fall results only from severe impairement of position and joint sense in the lower limbs. Patient show sensory ataxia while walking, being unaware of the position of his feet in relation to the ground. He walks with high stepping gait.

Ataxia of sensory[7] type may also be apparent in upper limbs if similarly affected the hands are clumsy and fine movements cannot be performed particularly when the hands are out of sight (useless hand syndrome).

1.Peripheral nerve lesions:

Along with general features of sensory ataxia, peripheral nerve lesions specifically show some additional features. They are glove and stocking type of sensory disturbance, all modalities of sensation, are impaired and associated signs of lowermotor neuron signs (wasting, hypotonia and hyporeflexia).

2. Posterior Nerve Root Lesions

1. Post Infective Demyelination – GBS like syndrome (Tornero etal – 1997, case report).

2. Tabes Dorsalis

3. CIDP

Features – Sensory Ataxia, Depressed All Modalities of Sensation And Diminished or Absent Tendon Jerk.

3. Posterior column in spinal cord.

1.Compressive myelopathy 2.SCD

3.Pellagra 4.Advanced syringomyelia

5.Multiple sclerosis

Brainstem lesion:

Sensory abnormalities can be easily interpreted on anatomical basis. Complete hemisensory loss is present if the lesion is in the upper midbrain, while medullary lesion can give facial sensory involvement on one side with hemianasthesia on the trunk and limbs on the opposite side. A midbrain lesion involving the 3rd nerve palsy. With contralateral static tremor, hamianaesthesia (Benedikt's Syndrome).

Etiology - Brainstem - Tumor - Infarct / Haemorrahage.

- Demyelination - M.S.

Thalamic Lesion:

Contralateral hemianaesthesia and thalamic pain are the prominent features. Fisher (1965) described pure sensory stroke in a patient with thalamic lacunar infarct, who had contralateral hemianaesthesia.

Etiology – Infarct / hemorrhage - Tumor

Sensory cortex lesion:

If sensory cortical lesion is irritative in nature it can cause sensory jacksonian, epilepsy. When there is destruction of part of post central gyrus, appreciation of position tactile discrimination, localization of size, shape and texture are impaired.

Etiology - ICSOL - Infarct - Hemorrhage.

VERTEGENOUS ATAXIA

Neurological Causes of Vertigo

Vertigo[54] is a very specific condition in which the environment or the patient himself seems of rotate. It is purely subjective, but may be associated with objective signs Acute Vertigo that lasts for days results from a unilateral loss of vestibular function. The loss can be from a peripheral lesion (lanyrinth or vestibular nerve) or from a central lesion (brain-stem or cerebellum).

Table -1

Difference Between Peripheral And Central Vertigo[54]

Symptoms	Peripheral Vertigo	Central Vertigo
Nausea, Vomiting	Severe	Moderate
Imbalance	Mild	Severe
Hearing loss	Common	Rare
Oscillopsia	Mild	Severe
Neurologic Symptoms	Rare	Common
Comprehension	Rapid	Slow

Spontaneous nystagmus of peripheral origin does not change in direction with gaze to either side, although it increases in amplitude with gaze in the direction of the fast phase (Alexander's Law)[54]. In contrast, spontaneous nystagmus of central origin typically changes direction when the patient looks away form the direction of the fast phase.

Infarction of the labyrinth, brain-stem or cerebellum typically occurs in older patient population with known vascular risk factors. Since the circulation to the labyrinth originates from the vertebro-basilar system, infarction of the labyrinth can be part of a larger brain stroke syndrome. For example with occlusion of the anterior inferior cerebellar artery (AICA) (Oas JG

1992)[5] labyrinthine infarction is commonly associated with infarction of lateral pontomedullary region and the anterior inferior cerebellum.

Vertebrobasilar insufficiency (VBI) (Gomez CR, Cruz – Floreesa, 1996)[4] is a common cause of spontaneous attacks of vertigo in older patients. Vertigo occurs in approximately 25% of unselected migraine patients. (Bickerstaff ER – 1961)

AIM OF THE STUDY

1. To study the various aspects of acquired and hereditary ataxic syndromes

2. To study about the level of involvement of the neuraxis in various type of ataxic syndromes.

3. Correlative study of CT scans finding and clinical features in acquired and hereditary ataxic syndromes.

MATERIALS AND METHODS

70 Patients were studied who studied who presented with ataxia as the main complaints. The period of study was from July 2004 to Jan 2006. All the cases were inpatients of medical and neurology wards or who attended neurology or ENT OPD in Coimbatore Medical College hospital. All the patients who had ataxia were chosen like cerebellar, sensory and vertiginous ataxia. Ataxia of acute subacute and chronic type and ataxia of hereditary and acquired type were also included in the study. Among the acquired type of ataxia due to stroke, infection, neoplasm, demyelinations, degeneration, drugs, alcohol and paraneoplastic syndrome were included in the study. Ataxia due to congenital bony anomalies like cranio vertebral junction anomaly was also considered. All patients had a detailed clinical work up which included history general examination and systemic examinations and relevant investigations to identify anatomical locations and nature of pathology leading to ataxia.

Demographic informations such as age, sex, occupations and percapita income were obtained. Details of social habits, such as alcohol consumption, smoking habits were enquired into. Exposure to STD, TB, Toxins, vaccine were also searched. Apart from the main complaint ataxic symptoms referable to cranial nerves, pyramidal system, extrapyramidal system, autonomic nervous system and also symptoms of raised. ICT were enquired into. Note was made on trauma, fever, exanthems, csom, drugs especially antiepileptics ATT and dapsone, etc.

Past history of SHT, DM, TB,RHD, STROKE were carefully enquired into Family history of ataxia was also searched. Details of consangunity also was obtained.

In the general examination, specifically looked for neurocutaneous markers, height neck ratio, anaemia, clubbing, lymphadenopathy, peripheral nerve thickening and any abnormality of peripheral vessels.

In higher function examination, more stress were given for speech and tried to find out type of abnormality like, scanning, staccato or spastic when it was present.

Individual cranial nerves were thoroughly examined and fundus was looked for optic atrophy papilleodema and pigmentary change, Nystagmus was thoroughly examined to findout whether it was due to central / peripheral lesions.

During motor system examinations, bulk, tone strength and deep tendon reflex, co-ordination and involuntary movements were looked to find out associated pyramidal, LMN, extrapyramidal and cerebellar lesion. When incoordination was present all possible clinical tests were done to findout whether it was due to cerebellar, sensory or vestibular abnormality. If the incoordination was due to cerebellar lesions, further clinical examinations was directed to find out the ataxia is limb ataxia or gait ataxia or truncal ataxia.

Sensory systems also thoroughly examined to find out which tract was involved and the site of lesion.Spine and neck examined to find out any feature of short neck and vertebral anomaly.

Other systems were also examined to findout any pathology for the patients problem eg., malignancy for paraneoplastic cerebellar degeneration and cardio vascular system to rule out cardiac cause of embolism.

PATIENTS SELECTION

Patients were broadly divided into three groups.

1. Ataxia of hereditary cause and 2. Ataxia of acquired cause and 3. Ataxia due to congenital bony anomaly where ataxia developed later on. Individual patients were studied accordingly to anatomy of the disease, onset of illness and pathology of the disease.

According to anatomy

1.Cerebellar ataxia 2.Sensory ataxia 3.Vertigenous ataxia

According to onset

Acute Subacute Chronic

According to Pathology

Vascular	Infection
Demyelinations	Degenerative
Tumor	Paraneoplastic
Drugs, toxins, alcohols	Trauma

Criteria for selecting patients with ataxia of cerebellar type

1. Limb, gait or truncal ataxia 2. Scanning or staccato speech.

3. Dysmetria → Intentions tremor 4. Dysdiadokokinesia

5. Coarse gaze evoked, horizontal nystagmus with fast component to same side of lesions. 7. Broad based gait.

Criteria for selecting patients with ataxia of sensory type.

General Features:

1. History of pins and needle with tingling sensations
2. Cotton wool sensations during walking.
3. Ataxia – swaying forward and backward.
4. High stepping gait
5. Rombergism

Peripheral nerve lesion producing ataxia

6. Hypotonia
7. Hyporeflexia
8. Glove and stocking sensory disturbance
9. Peripheral nerve thickening

Posterior columns cervical cord level.

10. Lhermitte sign

Due to medial leminicus.

- Other long tract sign - Cranial nerves

Criteria for selecting patients with ataxia of vertiginous type.

1. Vertigo 2. Tinnitus

3 Deafness 4. Nystagmus

- Fine gaze evoked with fast component to opposite side of lesion with some rotatory component. Werdelins in 1986 put forward some criteria for diagnosis of hereditary cerebellar ataxia syndrome. The same criteria were applied in the present study for diagnosing hereditary ataxic syndrome.

1. Signs of cerebellar dysfunctions

a. Ataxia of the extremities independent of vision.

b. Dysarthira – Scanning staccato

c. Dysmetria (Intention tremor)

d.Nystagmus

e. Hypotonia.

f. Broad based gait

2. Signs of pyramidal dysfunction

a. Paresis b. Babinski's sign

3. Amyotrophy

4. Deformities, usually club foot

5. Involuntary chorea – like movements

6. Extrapyramidal signs

a. Rigidity b. Bradykinesia

7. Optic atrophy

8. Dementia

9. Epilepsy

10. Retinal pigmentary changes.

Hereditary ataxia are again divided into early onset (before 20 years) and late onset. (After 20 years) Patients who are having a known etiology like infarct, haemorrhage, tumour, infection, degeneration, demyelination, trauma were taken as acquired ataxia in the present study. Investigations were done in detail for such patients and showed positively in most of the patients eg.1. If ataxia in one patients is suspected due to cerebellar neoplasm CT scan of brain taken and confirmed the diagnosis. 2. If ataxia is suspected due to demyelination of posterior nerve root (GBS) CSF analysis done to confirm the diagnosis.

Routine urine and blood investigations were done to find out any evidence of diabetes, infections and raised ESR. Special blood test like, VDRL thyroid functions test and liver functions test were done whenever appropriate.

X-ray chest PA view taken to rule out any foci of pulmonary tuberculosis, to rule out evidence of cardiology (RHD) and search for bronchogenic, pleural or mediastinal malignancy. X-ray of skull and cervical spine and were taken in suspected cases of CVJ anomalies and cervical cord compression.

CT scan of brain also was done (48 – cases) whenever appropriate to help the diagnosis. MRI scan was done when it was very essential to pickup up brain stem pathology and CVJ anomly. Lumbar Puncture and CSF analysis were done whenever appropriate like demyelination (GBS, CIDP).

Audiogram was done when ataxia was due to vertiginous cause. Caloric test could not be done in our hospital because of lack of facilities.

70 cases of various type of ataxia syndromes were studied. The various clinical syndrome encountered in the study were. (Table-1) Cerebellar ataxia 60.(85.7%), sensory ataxia 8(11.4%) and labyrinthine ataxia 2(2.8%)

Age Incidence

(Table- II) The age of the patients varied from 6 to 70 years. Lowest age noticed is 6 years who was suffered from Brain stem glioma. Highest age was 65 years who was having lateral medullary syndrome. Between 0-10 years there were 3 cases (4.2%) between 11.20 years 2 cases (2.8%) between 21-30 years 20 cases (28.5%) between 31-40 years, 17 cases (24.2%) between 41-50 years 13 cases (11.5%) between 51-60 years 10 cases (14.2%) and between 61-70 5 cases (7.1%) . The least incidence at 11-20 years period and maximum incidence 21-30 years period.

SEX INCIDENCE

Out of 70 cases men were 43 and women were 27. The ratio is 1.59:1

Main anatomical site of pathology:

(Table – III) Various anatomical site of involvement could be made out which leads to ataxic syndromes. They were pure cerebellum 26 cases (37.1%) leading the highest group, Then brainstem, 18 cases (25.7%), combined spinal cord and cerebellum 15 cases (21.4%), pure spinal cord 3 cases (4.2%), posterior root 3 cases (4.2%), labyrinth 2 cases (2.8%), peripheral nerve 2 cases (2.8%) and thalamus one case (1.4%)

Incidence of hereditary and acquired ataxia:

(Table IV) Out of 70 cases studied 50 (71.4%), were of acquired variety and 17 (24.2%), were of hereditary type. The congenital anomaly later on leading to ataxia were 3 cases (4.2%).

Among the 50 cases of acquired males were 31 and female were 19 ratio is 1.6:1. Among the 17 cases of hereditary ataxia 12 and 5 females Ratio is 2.4:1

Incidence of Hereditary type of Cerebellar and hereditary type of sensory ataxia:

(Table V) Hereditary cerebellar ataxia were 16 out of 17 hereditary ataxia and ataxia due to hereditary sensory neuropathy was 1 out of 17.

Age incidence of hereditary ataxia

There were totally 17 hereditary ataxia comprising both hereditary cerebellar and hereditary sensory ataxias. Between 0-10 years there was only one case. Between 21-30 there were 8 cases between 31-40 years, 5 cases, between 41-50 years 3 cases between 41-70 there were no cases. The congenital anomaly later on leading to ataxia (CVJ) present in this study was at the age interval of 31-60 years.

Incidence of various type of spinocerebellar ataxia

(Table – VI) In the present study there were 16 cases of spinocerebellar ataxia Among that OPCA(SCAI) was the major group of spinocerebellar ataxia which were 8 out of 16 cases SCA – II was 3, SCA – III – 1, there was no SCA 5 to 22. In the case of DRPLA, Friedriech's ataxia and ataxia telengiectasia one case of each was present.

Age and Sex incidence Hereditary Cerebellar Ataxia

(Table VII) There were 16 cases of Hereditary Cerebellar Ataxia in the present study. Between 0-10 years age there were one case. Between 21-30 years 7 cases, between 31-40 years 4 cases, between 41-50 years 4 cases. Between 11-20 years and 51 -70 years there were no cases of

hereditary cerebellar ataxia in the present study. In the present study there were 11 cases of male and 5 cases of female SCA present. Sex ratio was 2.2:1

Age and Sex incidence of OPCA.

(Table –VIII) In the present study out of 16 cases of SCA there were 8 cases of OPCA. Between 0-20 years there was no OPCA present in the present study. Between 21-30 years, there was 2 cases, between 31-40 years. 3 cases and between 41-50 years 3 case were present there were 4 males 4 females. The sex ratio was 1:1.

Clinical Features of acquired cerebellar ataxia:

(Table – IX) Ataxia of gait was the chief presenting symptoms in all the 44(100%) cases of acquired cerebellar ataxia. The youngest age of onset of symptoms was 8 years and oldest 65 years. The duration of illness varied from 1 to 10 years. Along with cerebellar ataxia 8 patients had sensory ataxia, 5 of which were due to posterior column involvement and 3 due to peripheral neuropathy.

The next common symptoms were dysarthira. 16 out of 44 patients had dysarthria as their second major symptom but examination revealed that 19 patients (43%) had dysarthria, 25 patients had normal speech. Among the 19 dysarthria patient, 15 had scanning type of speech and 4 had staccato speech. Dysarthria started simultaneously with difficulty in walking in 7 patients. Muscular weakness was a major symptom in 10 patients. But on clinical examination 16 patients had features of pyramidal involvement. (36.3%)

Wasting was present in 3 cases. But no fasciculations. Wasting was seen over deltoid infraspinatus and supraspinatus muscle bilaterally. 6 Patients complained of visual impairment. Out of 6,2 had mature cataract and 4 had immature cataract.

Fundus of 3 patients (6.8%) showed papilloedema and all of them had intracranial space occupying lesions. No patients showed optic atrophy. Deafiness was a major problem in one patient (2.2%) and on examination the deafness was of sensory neural type.

13 patients complained of difficulty in swallowing. However 18 patients (40.9%) were shown to have bilateral palatal palsy (9^{th} and 10^{th} N). In addition 5 patients (11.3%) had tongue wasting, autonomic symptoms were not present in any of the patients, But one patient showed features of hroner's syndrome Bladder was not involved in any of the cases.

Co-existing Hansen's disease was seen in one patient and CSOM in another 2 patients. 5 patients had evidence of ischemic heart disease 2 of them had inferior wall myocardial infarction and 3 had ischemic changes in ECG.

None of the patient had dementia. Gaze palsy was present in 6 cases, 2 of them vertical gazepalsy and 4 with horizontal gaze palsy. One had one and half syndrome and another had internuclear ophthalmoplegia. 6(13.6%) patients had facial palsy 2 of them had facial diplegia of LMN type. Two had UMN type of facial palsy on one side and 2 had unilateral LMN facial palsy.

None of the patients had any involuntary movement in the present study. Clinical examination showed lateral spino thalamic tract involvement in 19 cases (43.%). Kyphosis and scoliosis present in 3 cases (6.8%)

Family history of hereditary ataxia

A definite family history was obtained in 4 cases only in H.SCA. One patient who is a 35 year old lady with OPCA had an elder sister afflicted with the same symptoms. She could not be examined but the history obtained from the close relatives was in favour of hereditary cereballar

ataxia. The patient was born of a second degree consanguineous marriage. Parents did not show evidents of the disease.

Second patient, a 45years old male with positive family history, was a case of dentato rubropalidean leucean atrophy. Who had 6 siblings of these one had similar disease characterized by severe gait ataxia abnormal movements deafness and impaired memory. There was no history of consanguinity. Another patient a 29 years old male, who presented with SCA –Type II gave history of progressive ataxia in his uncle since 33 years. He had an younger brother who died of some unknown disease at the age of 15 years. He was born of non-consanguinous marriage. Another patient 25 year male presented with SCA 2 had elder brother with progressive ataxia.

Fifth patient, 38 years female who was a case of hereditary motor sensory neuropathy gave a family history of similar illness in one of her elder brother . Both of them were born of second degree consanguineous marriage.

Clinical Features of hereditary cerebellar ataxia

(Table – X) There was totally 16 cases of hereditary cerebellar ataxia in the persent study. Ataxia of gait was the main presenting complaint in all the 16 cases (100%). The youngest age of onset of symptoms was 10 years, who was a case of ataxia telengectasia and the oldest 45 years. The duration of these varied from 2 to 18 years, Sensory ataxia present in 2 cases (12.5%) which were due to peripheral neuropathy.

The second common symptom was dysarthria 10(62.5%) out of 16 patients had dysarthria as their next major symptom, Slurring dysarthria was present in 5 cases, 3 patients had scanning type of dysarthira and 2 patients with staccato speech.

One of the patients gave a history of generalized toxic clonic seizures from the age of 20 years on wards. 6 patients showed abnormal involuntary movements. One had choreoathetosis (6.25%). One had myoclonus (6.25%) one (6.25%) had fasciculations of muscle and 3 (18.75%) another had postural tremors.

Pyramidal weakness was present in 9 patients (56.25%). Only one patient (6.25%) of 16 had distal amyotrophy. In them there was bilateral symmetrical wasting of both upper and lower limbs on the distal parts.

2 patients reported visual impairments (12.%) . In one (6.25%) it was due to bilateral primary optic atrophy and in another (6.25%) was due to bilateral immature cataract. There was no ptosis in any of the patient (6.25%)

3 patients had slow saccades (18.75%) 2 patients complained of difficulty in swallowing. However 3 patients (18.75%) were found to have bilateral IX and X cranial nerve palsy. One patient had wasting of tongue.

Bladder disturbance in the form of hesitancy was reported by one patient (6.25%) . Co-existant pulmonary tuberclosis was found in one patient (X-ray chest PA view).

Out of 16 cases one patient (6.25%) had scoliosis and another patient (6.25%) had pescavus. No scleral or iridial abnormalities were present. No Keyser – Fleischer ring observed. One patient had (6.25%) dementia. Two had (12.5%) subnormal intelligence and one (6.25%) had gross mental retardation.

No abnormalities in smell were observed in any patients. Impairment of color sense was not seen in any patient. Retinal pigmentary changes were also not seen in any patient. 2(12.50%)

patients had normal muscle tone, another 7 had hypotonia and 3 had hypertonia, Among the patient with hypertonia 2 had spasticity and one had cogwheel rigidity,

Signs of cerebellar dysfunction were present in all 16 cases. Gaze evoked nystagmus to the point of fixation was observed in 7 cases. Finger nose incor-ordination was present in all (100%). 9 patients had extrapyramidal features (56.25%).

Computerized axial tomographic scan were taken in 10 patient out of 16. Six patient it was not affordable. Out of 10 scans, 9 showed (90%) bilateral cerebellar atrophy. One report showed a normal study (10%). 3 patients had general cerebral atrophy (30%).

Hereditary Cerebellar ataxia- Age and onset

(Table – XI) There were a total number of 16 hereditary cerebellar ataxias. Among this 2 were of early onset type whose symptoms started before 20 years of age. They were cases of Friedreich's ataxia and ataxia telengiciectasia. 14 were of late onset type, whose symptoms started after 20 years of age. Early onset type of spinocerebellar ataxia are autosomal recessive and late onset of autosomal dominant type.

Sensory Ataxia Incidence of site of lesions

(Table - XII) There were 8 patients with sensory ataxia in the present study 4 (50%) of them had lesion of posterior root, 2(25%) with lesion at the level of posterior columns and 2(25%) were with peripheral neuropathy.

Labyrinthine ataxia incidence

(Table XIII) In the present study 2 patients were with labyrinthine type of ataxia Both of them had peripheral labyrinthine lesions which lead to ataxia.

Acquired ataxia-Etiology

(Table XV,XVI) In the present study there were 50 patients with acquired ataxia. On analyzing the possible etiology, by doing various specific investigation like CT scan brain, MRI, X-ray spine, CSF analysis, nerve conduction study, and audiogram showed most of the clinical diagnosis were correct. By correlating the clinical diagnosis and specific investigations various causes of ataxia were obtained. Among these, vascular events were 22(44%) and which was the most common cause of acquired ataxia. Next common cause was neoplastic which were 7 patients (14.1%). Third one was infection constituting 6 patients (12%). There were 5 patients with degenerative cause (10%)4 patients with drug toxicity with antiepileptic drugs (8%)4 patients with demyelinative conditions and 1 Patient with paraneoplastic syndrome and 1 patient with traumatic cause.

On analyzing the vascular causes of ataxia different anatomical locations and different types of vascular accidents obtained. There were 6 cases of cerebellar stroke 8 patients with brainstem stroke, 5 patients with lateral medullary syndrome, 1 patients with thalamic infarct.

Among the neoplastic cause, 4 patients had cerebellar tumor,2patients with brain stem tumor and one patient with cerebello pontine angle tumour.

With the degenerative causes 2 patients were with alcoholic cerebellar degeneration and 1 patient with alcoholic peripheral neuropathy. Among the demyelinating conditions there were 2 patient with Guillain barry like syndrome 1 patient with CIDP and one patient with brainstem demyelination.

Among the infective causes 3 patients had cerebellar abscesses 2 patients acute labyrinthitis and one patient cerebellitis caused by typhoid fever. Apart from these group of patients there was 1 patient with paraneoplastic syndrome and 3 patients with spinal cord compression.

INVESTIGATIONS

CT Scan

CAT scan was taken in 48 patients (68.5%) and 35 patients showed various findings which lead to acquired ataxia and 9 patients showed features fitting for HCA and 4 were reported as normal study. CSF analays done for 6 patients out of which 3 patients showed albumino cytological dissociation.

Nerve conduction study were done in 2 patients (2.8%) and both showed peripheral neuropathic pattern. MRI was done for 7 patients (10%) one patient showed pontine glioma and the other patient showed Arnold chiary malformation. 2 showed cerebellar infarcts 1 showed cp angle tumour 1 showed cerebellar atrophy 1 showed brainstem demyelination.

Audiograms done for 2(2.86%) patients with acute labyrinthitis and showed normal study. Myelogram was done for 2(2.86%) patients who had suspected cord compression.

X-ray spine was taken for 6 patients. 2 patients showed cervical spondylitic change and one showed traumatic fracture of vertebra.

DISCUSSION

Clinical features of 70 cases of ataxic syndrome were studied in the course of 18 months. The different modes of clinical presentation were thoroughly studies and family members were examined as far as possible and specific investigations were done to find out possible etiology for the ataxic syndromes. Analysis of the patient series showed that acquired ataxic syndrome accounts for about 71.4% forming the largest clinical type of ataxic syndrome in this study. The rest of the series comprised 22.8% hereditary cerebellar ataxia, 1.4 % hereditary sensory neuropathy and 4.2% with congenital causes which later on lead to ataxis syndrome (CVJ anomaly).

The syndrome of cerebellar ataxia 85.7% formed the commonest clinical type of ataxic syndrome in this study. Sensory ataxic syndrome were of 11.4% and labyrinthine ataxic syndrome(2.8%).

Etiology wise, cerebrovascular accident were the most common cause of acquired ataxic syndrome 44% second commonest cause was neoplasia 14%. The other causes of acquired ataxic syndromes in this study were infection 12% degenerative conditions, 10% drugoxicity 8% demyelinating diseases 8% paraneoplastic syndrome 2.1% and trauma 21%.

Main anatomical site of pathology of the ataxic syndrome were also studied in the present study. Pure cerebellum is the major anatomic lacation of the pathology, 37.1% in this series of study. The second commonest site of pathology is brain stem 25.7% and 21.47 other sites of pathology in this study are pure spinal cord 4.2% posterior root 4.2% peripheral nerve 2.8% and labryrinth 2.8%.

Young age of ataxic syndrome due to cerebellar involment in the present study considering both acquired and hereditary was 6 years and oldest age was 65 years. 1st patient was affected by

pontine gioma and 2nd patient with lateral medullary syndrome. The most common age group affected by cerebellar ataxic is 21- 50 years. The incidence of cerebellar ataxic syndrome is less between the age group 0-10 and between 51 and 70 years in the present study.

Similar to cerebellar ataxic syndrome the most common age group affected by sensory ataxic syndrome is 21-50 years. No cases of sensory ataxic syndrome present between 0 to 10 years and between 60-70 years in the present study. Vertegenous ataxic is common between the age group 41-60 years in this series of study.

Out of 70 cases ataxic syndromes men are 61.4% and women are 38.5% with ratio of 1.6:1. Acquired and hereditary ataxic were more common in male compared to female in the present study.

ACQUIRED ATAXIA

Acquired ataxic syndrome formed the commonest clinical type of ataxia in the present study 71.4%. the mean age of symptomatic presentation was 45 years. The youngest age of onset was 6 years , pontine glioma syndrome and oldest age was 65 years with lateral medullary syndrome . Within the acquired ataxic syndrome cerebellar ataxia are 88% sensory 14% and labyrinthine 4%.

Ataxia of gait independent of vision was present in all cases of acquired cerebellar ataxia 100% -It was the first symptom in all cases except 4. In these 4 patients, deafness and tinnitus was the commonest symptom. Among these , one patient was diagnosed as a case of acoustic neuroma. Other two patients who presented with occipital headache later on proved to have cerebellar tumors. The 4th patient who presented with generalized tiredness and loss of weight turned out to be a case of para neoplastic syndrome. The 44 patients of acquired cerebellar ataxia, 3 patients belong to the group congenital anomaly which later on developed ataxia (CVJA). 19 patients had

dysarthria 43% Extrapyramidal signs were not present in any of the acquired cerebellar ataxic syndrome. Pyramidal tract signs were present in 16 patients 36.3% posterior column involvement was seen in 11.35 papilloedema was observed in 3 patients 6.8% but none of the patients showed optic atropy. Skeletal deformities like kyphosis and scoliosis was present in 3 cases (6.8%). Dementia, mental retardation or subnormal intelligence were not noted in any of the patients . No patient had seizure. Partial ptosis seen in one patient which was due to Horners syndrome due to brain stem glioma. Gaize palsy seen in 6 patient 13.6%. Among the gaze palsy both horizontal and vertical gaze palsy were seen. One patient had one half syndrome with ataxic nystagmus, which was a case of brain stem infarct. Sensory neural deafness was present in one case 2.2% which was a case of acoustic neuroma. Facial palsy was present in 6 patients 13.6% palatal palsy in 18 patients 40.9% tongue weaken in 5 patients 11.3%.

ACOUSTIC NEUROMA:

Harner S.G. [13] (1983)in his studies about acoustic neuroma, mentioned that the highest incidence of this tumour is in the sixth decade. In the patient study the patient was a female of age 48 years. Other features were also comparable to ojeman and co – workers study. In his study of acoustic neuroma, the earliest symptoms were deafness (33 patients), headache(4), and unsteadiness 3. In the present study patient symptoms were deafness, headache and unsteadiness.

Earliest Symptom	**Ojemans Study[57]**	**Present Study**
Deafness	33 out of 46	+
Headache	4 out of 46	+
Ataxia	3 out of 46	+
Facial pain	1 out of 46	-
Tinnitus	1 out of 46	+

Facial weakness	1 out of 46	+

Cerebellar Stroke

In the present study CAT scan showed cerebellar hemorrhage in 4 patients and cerebellar infarct in a 4 patients. Fuji moto M[3] et al in 1997 had done a study of clinical features of CT proved cerebellar stroke in 16 patients selected in neurology ward in seirei Hammatsu general hospital.

Symptoms	**Fujimoto study**	**Percentage**	**Present Study**	**Percentage**
Vertigo	14	87.5	5	83.3
Truncal Ataxia	9	56.2	4	66.6
Limbs ataxia	5	31	3	50
Gaze evoked nystagmus	5	31	4	66.6
Total No.of Cases	16		6	

In Fujimoto study out of 16 patients with cerebellar stroke, 14 (87.5%), had vertigo, in the present study it was 83.3%. Truncal atxia 9 patients (56.2%), limb ataxia patient (31%), and gaze evolved nystagmus was 5(31%) in Fujimoto study. Corresponding results were obtained in the presenting study also with truncal ataxia 66.6% limb ataxia 50%, nystagmus 66.6%.

Thalamic Infarct

Martinez P[2] and Carrera et al studied about various clinical features of bilateral thalamic infarct in 1997. They compared their observation with various studies obtained from literature and found that almost comparable to that results.

S.No.	Symptoms	Martinez Study	Present Study
1	Disorder of Consciousness	+	+
2	Oculomotor disorders	+	+
3	Cerebellar symptoms	+	+
4	Memory disorders	+	+
5	Sensory symptoms	+	+
6	Abnormal movements	+	-
7	Pyramidal sign	+	+

On comparing the Martinez study the present study showed resemblance to a great extent. Only one different finding was that, there was no case with abnormal movements at the present study, but it was present in the Martinez study. But to the present study there was only a single case i.e., bilateral thalamic infarct.

Ataxic Neuropathy

Harada K, Nagata H. et al[1] have done a study about etiology of 6 cases of ataxic neuropathy in adult in 1997 at baraki prefectual central hospital. The results of the study differ from the present study.

Etiology	Harada Study (Total 6)	Present Study (Total 3)

	No.of Cases	Percentage	No.of Cases	Percentage
Carcinoma	2	33.3	0	0
Sjogrens syndrome	1	16.6	0	0
CIDP	1	16.6	1	33.3
Chronic idiopathic ataxic neuropathy	2	33.3	0	0
Alcoholic peripheral neuropathy	0	0	1	33.3
Hereditary motor sensory neuropathy	0	0	1	33.3

In Harada's[1] study about etiology in 6 cases of peripheral neuropathy leading to ataxia, carcinoma 33.3%, sjogrens syndrome 16.6% CIDP 16.6% and chronic idiopathic ataxia, neuropathy 33.3%. Among those conditions, present study consist only of CIDP and is of 33.3%. In the present study alcoholic peripheral neuropathy 33.3% and hereditary motor sensory neuropathy is 33.3%.

Acquired sensory ataxis, were also a major group of patients of in the present study. There are 6 patients with acquired sensory ataxia in the present series among the sensory ataxia, posterior nerve root pathology heading the commonest i.e., 50% Other sites of pathology are peripheral nerve (25%) and posterior column (25%). In the present study no patients with sensory ataxia due to medial leminiscus lesion or thalamus lesion was found out.

There are 2 cases of labyrinthine ataxia (2.8%) in the present study among which both are due to peripheral labyrinthine pathology. For these patients audiogram was done, which showed normal result. Caloric test was not done because of absence of facility in the hospital.

HERIDITARY ATAXIA

There were 17 cases of (24.2%) ataxic syndrome due to hereditary causes obtained in the present study. In this group 16 cases were due to spinocerebellar degeneration and one case was due to hereditary motor sensory neuropathy.

Out of the 16 hereditary spinocerebellar ataxia, Olio ponto cerebellar autophy was the commonest one , which are 8 in number. Next common spino cerebellar ataxia was –SCA II. Each cases of spino cerebellar ataxia –III, VII, Dentato rubro palledeo lucean atrophy, Freidriech's ataxia and Ataxia telengectasia were obtained.

The mean age of onset of SCA was 28 years. Youngest age of onset was 8 years. The oldest age of onset was at 45 years. Most of the patents with SCA were in between 21 to 40 years. The male female ratio 2.2:1.

Ataxia of gait independent of vision was present in all cases (100%). It was the 1st symptom in 10 cases (83.3%) out of twelve. In other two patients involuntary movements preceeded the onset of ataxia. 10 patients had dysarthira(62.5%) Extrapyramidal signs were present in 6 pateints (37.5%). Chorea was present in one case and myoclonus in another. 3 had tremor, Pyramidal signs present in 9(56.25%) seizure in one case (6.25%), optic atrophy in one (6.25%) case, lower cranial nerve involvement in 3(8.75%). Amyotrophy in 1(6.25%), pes cavas in 1(6.25%) Dementia was seen in one case (6.255) slow saccades seen in 3 cases (18.75%).

Positive family history was present in 5 patients (31.2%). But a negative family history could not exclude subclinial disease in the family. When the data were analysed on the basis of the classification put forward by Harding (1983)[49] it was found that 2 patients (12.5%) in the present study belonged to the subgroup of early onset cerebellar ataxia (onset after 20 years).and 14 cases (87.5%) belong to subgroup of late onset cerebellar ataxia. (Onset after 20 years).

In her study Harding found that among the EOCA, 67.5% of cases were Friedreich's ataxia but in the present study it was found to be 50%. Other 50% of case was ataxia telengiectasia.

Virtually all tehdisorders of early onset are of autosomal recessive inheritance and the later onset ones are of autosomal doninat.[49]

Early cerebellar ataxia	Harding's study	Present study
Friedreich's ataxia	67.5%	50

Among the various type of late onset cerebellar ataxia, described by Harding(1983)[49] common type present in the present study was OPCA 8 (57.1%) charcterised by pyramidal extrapyramidal slow saccades, ataxia and optic disc pallor or optic atrophy.

Among the 8 cases of OPCA, definite family history was present only in one patient and the inheritance pattern of the disease was probably autosomal dominant.

Bercanio (1982) in his review of 117 cases of OPCA, came across 54 familial cases and 63 sporadic cases. In the present study only one patient was familial and rest of the 4 cases were sporadic(4).

	Familial OPCA	%	Sporadic OPCA	%
Present study	1	12.5	7	87.5
Bercanio's study	54	46.2	63	53.8

Bercanio[72] reported 46% autosomal dominant inheritance in familial OPCA and 54% were autosomal recessive pattern. Present study showed autosomal dominant pattern in the familial OPCA. He observed that familial cases began earlier and lasted longer than sporadic cases and abnormal movements and opthalmoplegia were more frequent in familial cases.

Age of Onset

	Familial	Sporadic

Present study	30 years	20-50 years
Bercanio study	2 months to 53 years	Congenital to 60 years

The age of onset of the disease ranged from 2 months to 53 years. In the present study it was 30 years for familial cases.

The initial symptoms was gait ataxia in the majority of cases in the study which was in accordance with the observation of Bercaino In this study dementia noted in 6.25% patients. Bercanio commented that dementia was a dominant feature of familial OPCA and that it might appear in any phase of the disease or especially in the middle to late period. The relative infrequency of dementia in the present study could be attributed to the short period of observations.

Pyramidal signs were present in 56.25% of the present series. This in full agreement with the observation made by Bercanio. Bladder disturbance was rare in the present series. One patient had urinary symptoms but he had prostatism. The observations was also comparable with Bercanio's report. He remarked that urinary incontinence developed very late in the course of illness. In Bercanio's study 51 patients had urinary incontinence. Of these 28 had dementia, 27 had posterior column dysfunction and 7 had pyramidal involvement. Primary optic atrophy was noted in 6.25% Bercanio results were comparable.

Lower cranial nerve involvement is 18.75% in the present study. Bercanio[14] (1982) said the lower cranial nerve involvement was a frequent observations in patients with OPCA and all of them presented with dysphagia. He did not give exact percentage.

In the report of 32 cases of hereditary ataxia by chuttani et al (1961)[26], all cases are sporadic. In the present study 87.5% are sporadic. Sarma and virmanis study (1971) had positive

family history in 8 cases. Youngest patient in their series was 16 years old and oldest 58 years. Skeletal deformities were reported in 24.7% cases.

Hoffman (1971)[32] in his report of hereditary late onset cerebellar degeneration gave description of 6 affected members of a family and all were of autosomal dominant patterns. All cases had onset in the third decade . A large group of 750 members were studied by Gurier (1972)[39] who stressed that late onset cerebellar degeneration were dominantly inherited heterogenous groups.

Sahadevan (1965)[38] from his study of hereditary ataxias in calicut noted a comparatively earlier onset of the disease 33 years. In the present study almost identical observations was made, 30 yrs. Wadia and Swami[64] in 1971 reported a family from Bombay with heredodamilial spinocerebellar degeneration. They observed slow saccadic eye movements in 16 cases, According to them slow saccade was a striking feature of an indigeneous veriety of hereditary ataxia in our country. In the present study slow saccade were observed on 3 patients.

Computerized axial tomographic scan

CAT scan was done in 48 cases of cerebellar ataxic syndrome. Out of which 10 were for hereditary ataxic syndrome and 38 for acquired cerebellar ataxic syndrome. For patients with hereditary ataxia CAT scan could not be taken, because it was not affordable. For all acquired cerebellar ataxic syndrome. CAT scan was taken whenever appropriate.

Out of 10 CAT scans of hereditary ataxia 1 showed normal study. 9 patients had abnormal scans. Salient abnormalities noted were atrophy of brain stem, cerebral peduncle and cerebellum. Marked atrophy of the cerebellum with prominent foliation and enlargement of fourth ventricle were seen in all abnormal scan. One cases showed diffuse cerebral atrophy. These findings are typically described in OPCA by previous workers.

Out of 38 CT scan of acquired cerebellar ataxic syndrome 2 showed normal study. The clinical diagnosis of the cases were lateral medullary syndrome and brainstem stroke. All other CAT scan showed definite findings favouring the clinical diagosis. The findings in CAT scan were infarct or hemorrhage in brain stem or cerebellum, tumor in cerebellum, cerebellar aneurysm, CP angle tumour, cerebellar atrophy, cerebellar abscess, tuberculoma in midbrain and cerebellar degeneration. Among the CT Scan reports of 5 cases of lateral medullary syndrome 2 showed additional cerebellar infarct.

Nerve conduction study were done in 2 sensory ataxia and showed pheripheral neuropathic findings. Audiogram done for all labyrinthine ataxia and showed normal reports. CSF analysis done in 6 cases. 3 showed features in favour of demyelination and one showed features of bacterial meningitis. X-ray skull APand lateral view, and open result in favour CVJ anomaly. Myelogram done for 2 cases of cord compression. MRI scan were done only for 7 cases one showed Arnold chiarry malformations and 1 showed pontine glioma 2 showed cerebellar infact, 1 showed cp angle tumour 1 showed cerebellar atrophy and 1 showed Brain stem demyelination. X-ray chest was taken whenever appropriate. One of the X-ray of patient with paraneoplastic cerebellar degeneration showed mediastinal widening and possibly that may be due to mediastinal tumour

Comparative incidence of Friedreich's ataxia in various studies.

S.No	Author	Year	No.of cases	Percentage
1	Chuttani et al	1961	27	84.3
2	Jolly	1961	9	45
3	Sahadevan[38]	1965	4	19
4	Chudhary st al	1968	4	57.1

5	Harding [48]	1983	157	65.5
6	Present study	2006	1	7.7

CONCLUSION

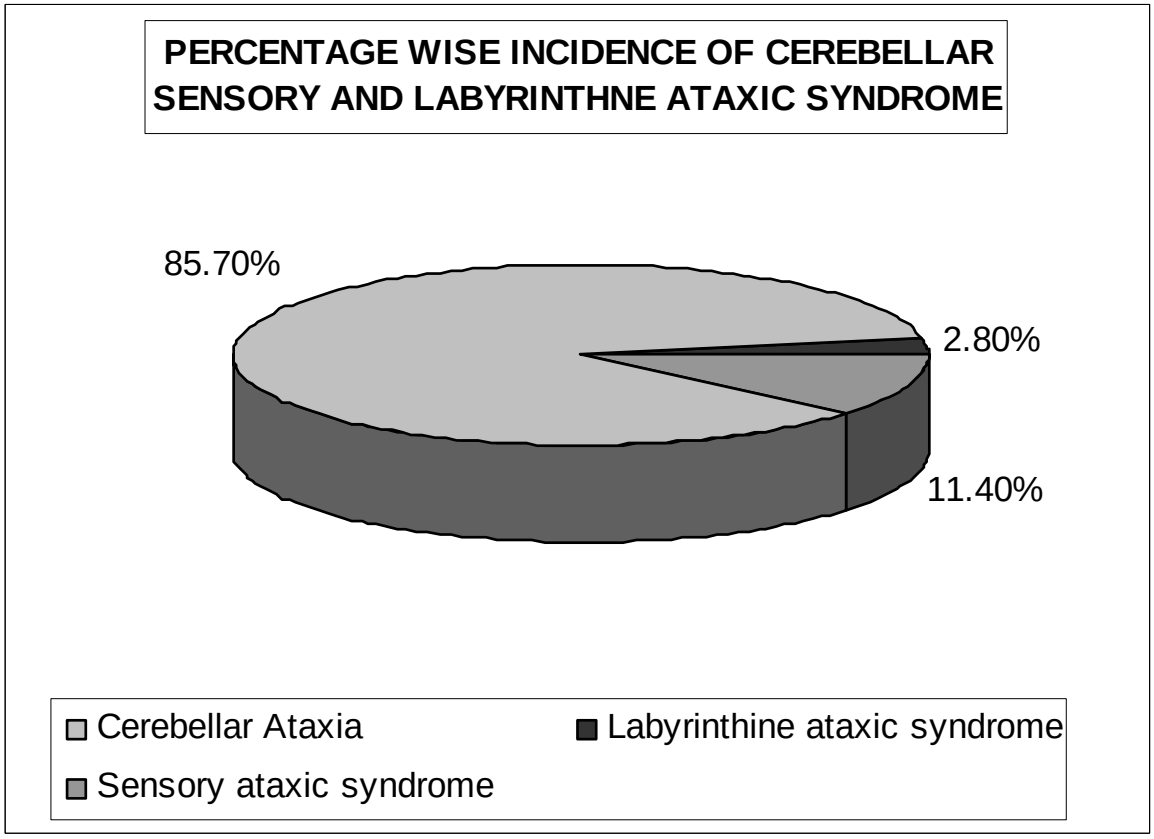
70 cases of various types of ataxia syndromes were studied during a period of 18 months. Their symptoms and signs were analysed. Cerebellar ataxia was 85.7%, Sensory ataxia 11.4% and labyrinthine ataxia 2.8% . The syndrome of acquired ataxia (71.4%) was the single major group encountered in this series. Among the Acquired ataxic syndrome, ataxia of cerebellar type is the predominant one (81.1%)and commonest cause of acquired cerebellar ataxic syndrome was vascular (44%.)

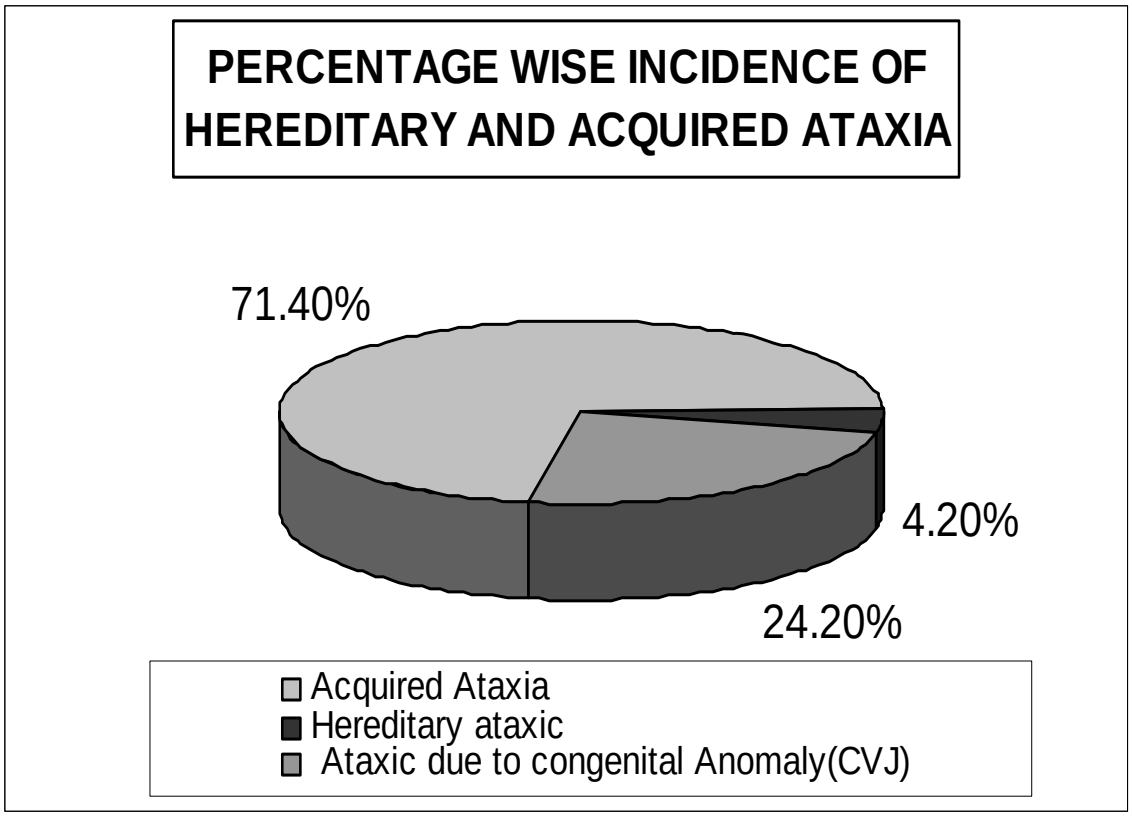
Next common cause of acquired cerebellar ataxic syndrome were neoplasms, infections, degeneration and drugs. Percentage of hereditary cerebellar axiac is 24.2% and OPCA were the commonest variety encountered (50%). The familial incidence (31.2%) of the hereditary ataxia was not found to be very variable on both acquired and hereditary ataxia was found to be less, 68.8% of the cases being sporadic.

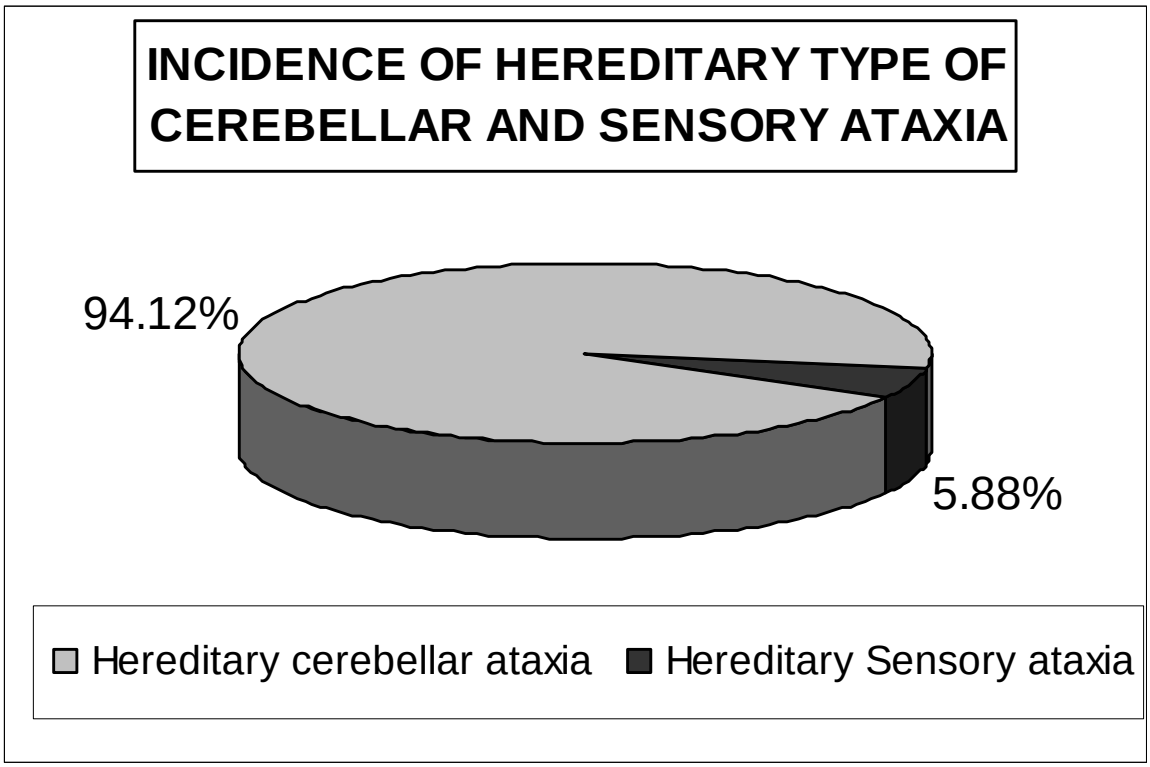
The clinical features were found to be very variable on both acquired and hereditary ataxic syndrome. Tomographic scan showed cerebellar and brain stem atrophy in hereditary cerebellar ataxia and for acquired cerebellar ataxia, showed vascular, neoplastic, degenerative, infective, changes.

SUMMARY

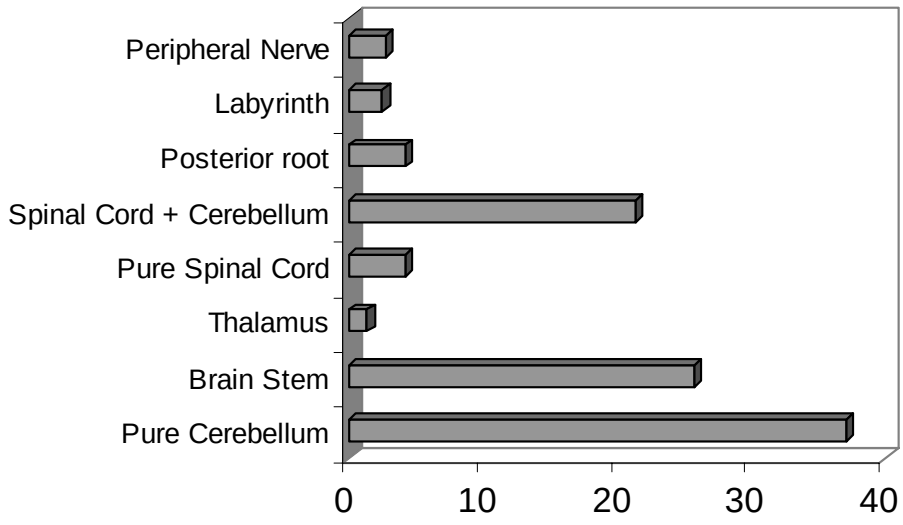
Friedreich was the fast person to gave chemical description of ataxia. Three types of Ataxias are of practical clinically important, they are sensory. Cerebellar and Vertegenous. Various studies on ataxia was done by Wadia, Chuttenn, Bercanio and Mautz. In persent study 70 patients were studied who presented with ataxia as main complaint. The study was done for 18 month period. During the study relevant investigation available in the hospital was utilized in establishing diagnosis. It was found out cerebellar ataxia was commonest type encountered in both acquired and hereditary groups and acquired form was more common than hereditary. Among cerebellar, vascular etiology topped the list. Second commonest type of ataxia encountered was sensory ataxia followed by labyrynthine ataxia . Among hereditary ataxia most of the cases were sporadic rather than familial.


PERCENTAGE WISE INCIDENCE OF CEREBELLAR SENSORY AND LABYRINTHNE ATAXIC SYNDROME
85.70%
2.80%
11.40%
Cerebellar Ataxia
Labyrinthine ataxic syndrome
Sensory ataxic syndrome

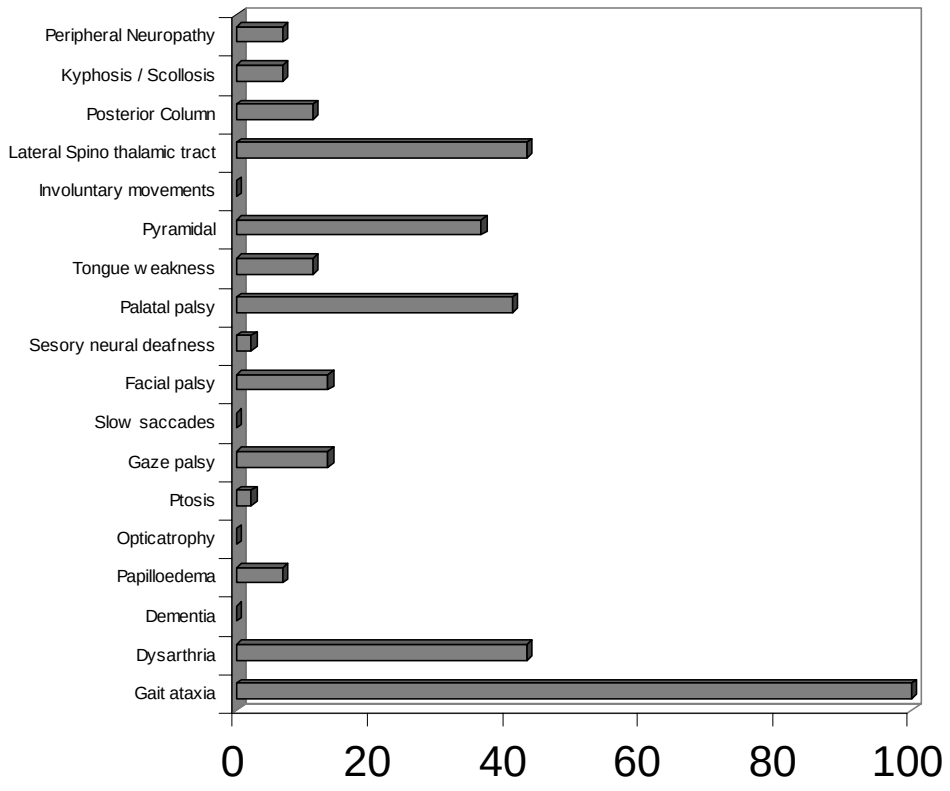

PERCENTAGE WISE INCIDENCE OF HEREDITARY AND ACQUIRED ATAXIA
71.40%
4.20%
24.20%
Acquired Ataxia
Hereditary ataxic
Ataxic due to congenital Anomaly(CVJ)


INCIDENCE OF HEREDITARY TYPE OF CEREBELLAR AND SENSORY ATAXIA
94.12%
5.88%
Hereditary cerebellar ataxia
Hereditary Sensory ataxia

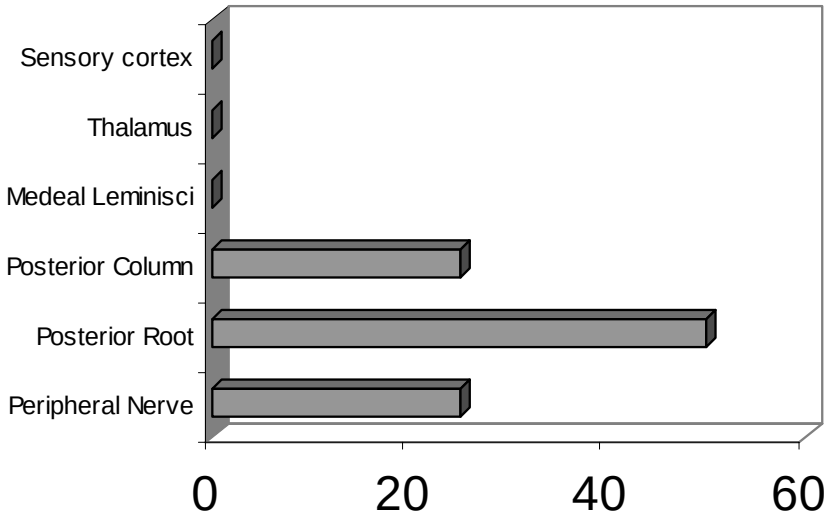
MAIN ANATOMY SITE OF PATHOLOGY

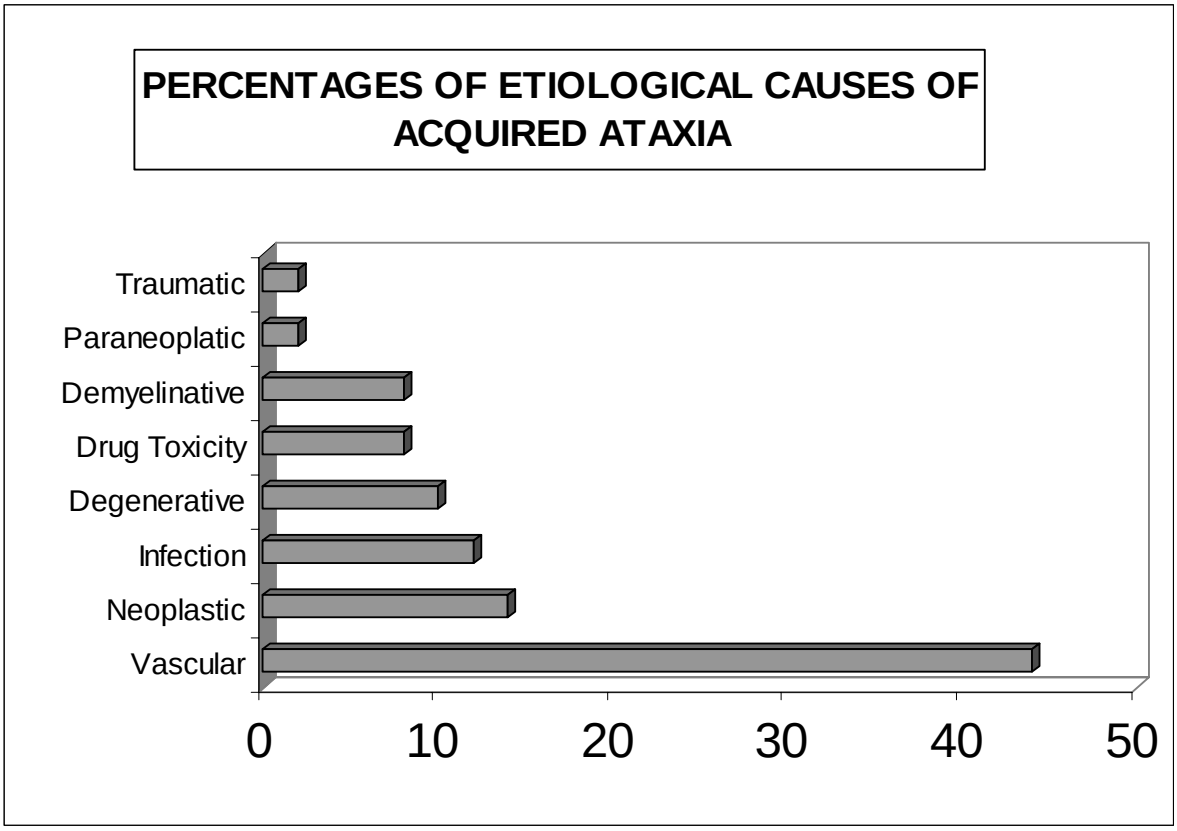


PERCENTAGE WISE CLINICAL FEATURES OF ACQUIRED CEREBELLAR ATAXIA

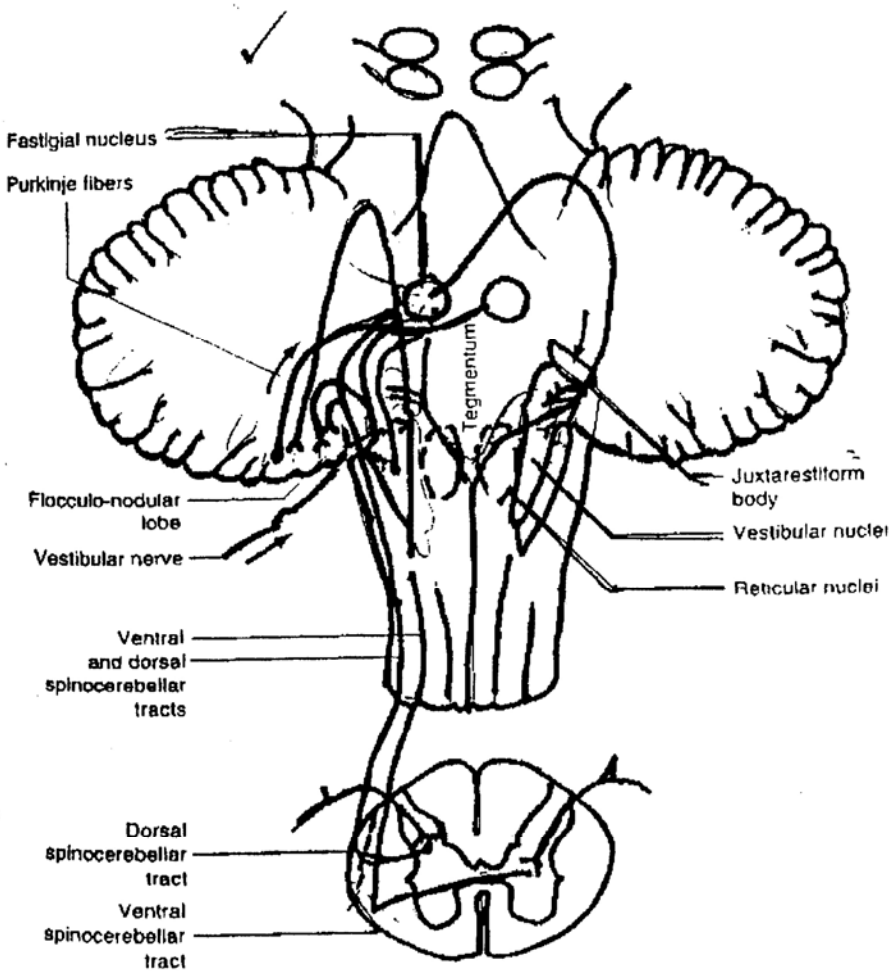


PERCENTAGE INCIDENCE OF SITE OF LESION IN SENSORY ATAXIA

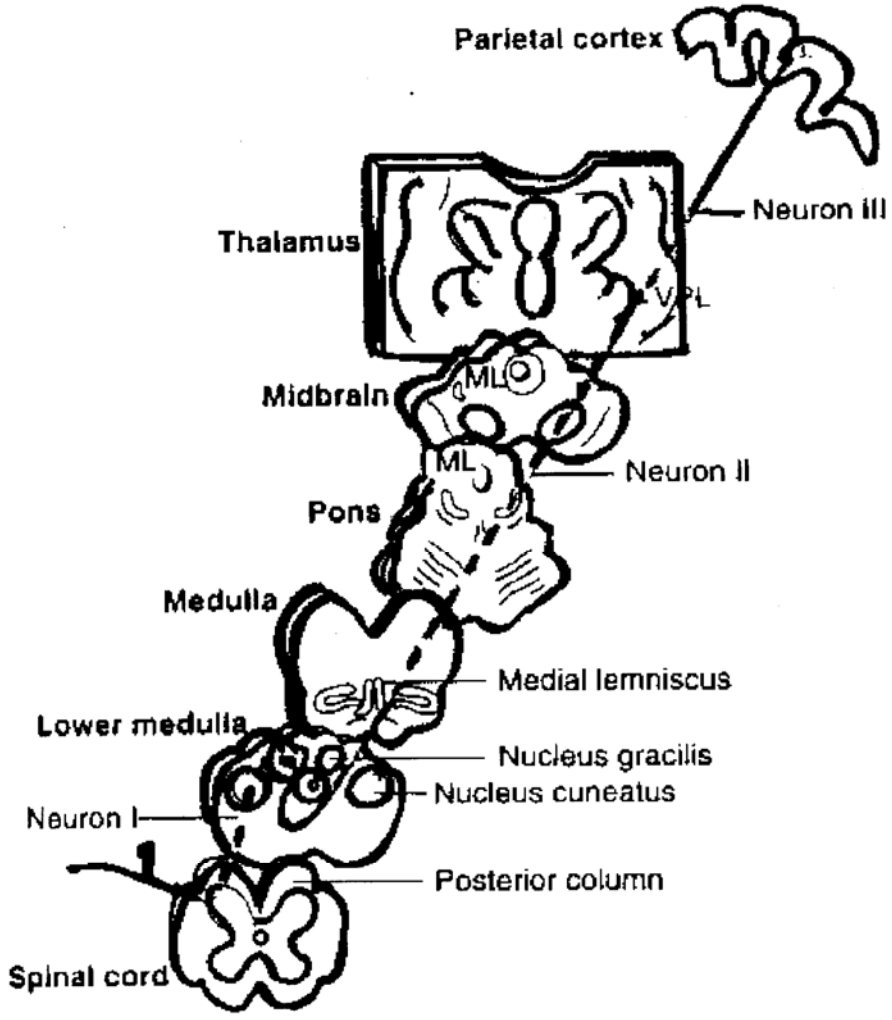



PERCENTAGES OF ETIOLOGICAL CAUSES OF ACQUIRED ATAXIA
Traumatic
Paraneoplatic
Demyelinative
Drug Toxicity
Degenerative
Infection
Neoplastic
Vascular
0
10
20
30
40
50

THE CONNECTIONS OF CEREBELLAM



POSTERIOR COLUMN PATHWAY



PROFORMA

Serial No.

Statistical data

Name Address Occupation

Age Sex:M/F

IP / OP No :

Symptoms

On set Acute Subacute Insedious

Swaying to either side YES / NO

Tremulousness of hand YES / NO

Numbness of lower limb YES / NO

Difficulty in walking in night YES / NO

Stiffness and Heaviness of limbs YES / NO

Tripping / Buckling YES / NO

Weakness of limbs YES / NO

Involuntary movements YES / NO

Bladder

Bowel

Disturbance of vision YES / NO

Diplopia YES / NO

Numbness of Face YES / NO

Difficulty in Chewing YES / NO

Facial Deviation towards Rt / Lt YES / NO

Deafness / Tinnitus YES / NO

Nasal Regurgitation			YES / NO
Signs of Raised Intracranial pressure			YES / NO
Trauma / Fever / Exanthems			YES / NO
Alcohol			YES / NO
Drugs			YES / NO
CSOM symptoms			YES / NO
Past History		HTN /DM / TB	
Family History		Relevant	YES / NO
General Examination			
Pulse Rate		BP	
Pallor			YES / NO
Neurocutaneous Markers			YES / NO
Height neck ratio			
Neurologic examination			
Higher functions			
Consciousness		Normal / Abnormal	
Orientation		Normal / Abnormal	
Intelligence		Normal / Abnormal	
Speech		Normal / Abnormal	
Cranial nerve			
Spinomotor system		RT	LT
Bulk	UL		
	LL		
Tone	UL		
	LL		

Strength	UL	
	LL	
DTR	UL	
	LL	
Sensory System		
Pain & Temp	UL	
	LL	
Vibration sense	UL	
	LL	
Position sense	UL	
	LL	
Cerebellar signs		
Head nodding	Yes / No	
Nystagmus	Yes / No	
Disdiadokokinesia		YES / NO
Rebound phenomenon		YES / NO
Pendular kneejerk		YES / NO
Tandem Walking		YES / NO

Gait – Normal – Spastic, - Ataxic – Stamping / Others

Extra Pyramidal signs YES / NO

Carotid Bruit RT LT

Skull & Spine Normal / Scoliosis / Kyphosis

Peripheral Nerve thickening YES / NO

Other Systems

Investigation:

Urine	ALB	Sugar	Deposit
Blood	TCL	DLC	HB – ESR

SUGAR UREA

T3 T4 TSH

SPECIAL BLOOD TESTS

ECG

RADIOGRAPHY

CT SCAN

MRI

LUMBAR PUNCTURE - CSF ANALYSIS

MASTER CHART

S. No	Name	IP No.	Age	Sex	Family H/O	Drug H/O			CSF Analysis	Myelogram	NCS	X-Ray		CT SCAN	MRI	Diagnosis
						Al	E	C				Chest	Skull & Spinal			
1	AJAY	5642	20	M	A	A	A	A	-	-	-	-	-	CAB	-	CAB
2	AMUDHA	4892	41	F	A	A	A	A	-	-	-	-	-	CA	-	SCA-I
3	ANNAPORANI	60651	56	F	A	A	A	A	-	-	-	-	-	CI	CI	CS
4	ARUMUGAM	3962	65	M	A	A	A	A	-	-	-	-	-	CI + BSI	-	LMS
5	AYYASAMY	56457	55	M	A	A	A	A	DM	-	-	-	N	-	-	GBS
6	BAGYAM	31942	35	F	P	A	A	A	-	-	-	-	-	-	-	SCA-I
7	BALAMANI	62931	43	F	A	A	A	A	-	-	-	-	-	-	-	SCA-I
8	BALASUBRAMANI	50616	29	M	A	P	A	A	BM	-	-	-	-	CAB	-	CAB
9	BALU	28944	30	M	A	A	A	A	-	-	-	-	-	CH	-	CS
10	BASKAR	62544	46	M	A	A	A	A	-	-	-	-	-	BSI +CI	-	LMS
11	CHANDRA	90648	13	F	A	A	A	A	-	-	-	-	-	CAB	-	CAB
12	CHANDRASEKAR	26224	45	M	P	A	A	A	-	-	-	-	-	-	CA	DRPLA
13	CHELLIAH	2962	42	M	A	A	A	A	-	-	-	-	-	-	-	LAS
14	CHELLAMMAL	8122	56	F	A	A	A	A	-	-	-	-	-	N	-	BSS
15	CHELLAMMAL	3692	40	F	A	A	A	A	-	-	-	-	-	BSI	-	LMS
16	CHITRA	89246	8	F	A	A	A	A	-	-	-	-	-	N	-	AT
17	DHANABAGYAM	36692	40	F	A	A	A	A	-	-	-	-	-	CH	-	CS
18	DHANALAKSHMI	28098	51	F	A	A	A	A	-	-	-	-	-	BSI	-	CMS
19	DHANALAKSHMI	76421	44	F	A	A	A	A	-	-	-	-	-	CA	-	SCA-I
20	DHANDAPANI	92564	33	M	A	A	A	A	-	-	-	-	-	CA	-	SCA-I

S. No	Name	IP No.	Age	Sex	Family H/O	Drug H/O			CSF Analysis	Myelogram	NCS	X-Ray		CT SCAN	MRI	Diagnosis
						Al	E	C				Chest	Skull & Spinal			
21	EZILAN	59465	29	M	A	A	A	A	-	-	-	-	-	CA	-	SCA – I
22	FATHIMA	8642	36	F	A	A	A	A	DM	-	-	-	N	-	-	GBS
23	GAYATHRI	5616	49	F	A	A	A	A	-	-	-	-	-	BSI	-	LMS
24	HEMA	2264	17	F	A	A	A	A	-	-	-	-	-	N	-	CVJ
25	HEMANATHAN	89465	25	M	A	A	A	A	-	-	-	-	-	CA	-	FA
26	IYAPPAN	36086	24	M	A	A	A	A	-	-	-	-	-	CA	-	SCA -4
27	JAMILABANU	46612	54	F	A	A	A	A	-	-	-	-	-	BTI	-	BTI
28	JASMINE	94268	38	F	P	A	A	A	-	-	PN	-	-	-	-	HSMN
29	JOHNSON	4283	47	M	A	P	A	A	-	-	-		-	BSI	-	BSS
30	KANNAMAL	6086	40	F	A	A	A	A	-	-	-	MM	N	CD	-	PNS
31	KARUPUSAMY	4122	52	M	A	P	A	A	-	-	-	-	-	CD	-	ACD
32	KRISHNASAMY	2695	48	M	A	P	A	A	-	-	-	-	-	PNET	-	CT
33	KUMAR	50896	29	M	P	A	A	A	-	-	-	-	-	-	-	SCA -2
34	LATHA	52210	48	F	A	A	A	A	-	-	-	-	-	-	AN	CPAT
35	MANIAMAL	94680	40	F	A	A	A	A	-	-	-	-	-	AC	-	CT
36	MARIMUTHU	93422	33	M	A	A	A	A	-	-	-	+	-	CA	-	SCA -1
37	MYLAPPAN	85638	44	M	A	A	A	A	DM	-	-	-	-	-	-	CIDP
38	NANDAKUMAR	50489	24	M	A	P	A	A	-	-	-	-	TFV	-	-	CC
39	NARAYANAN	12298	52	M	A	A	A	A	-	-	-	-	-	BSH	-	BSS
40	PALANIVEL	9246	46	M	A	A	p	A	-	-	-	-	-	-	-	ET

S. No	Name	IP No.	Age	Sex	Family H/O	Drug H/O			CSF Analysis	Myelogram	NCS	X-Ray		CT SCAN	MRI	Diagnosis
						Al	E	C				Chest	Skull & Spinal			
61	THANGAMANI	6042	49	M	A	P	A	A	-	-	-	-	-	CD	-	ACD
62	THILAGAVATHI	8221	6	F	A	A	A	A	-	-	-	-	-	PG	PG	BST
63	VADIVU	962	49	F	A	A	A	A	-	-	-	-	-	BSI	-	BSS
64	VEERAKUMAR	9048	28	M	A	A	A	A	-	-	-	-	-	-	-	SCA-1
65	VELLAPPAN	2606	29	M	A	A	A	A	N	-	-	-	-	BSD	-	BSD
66	VELLINGRI	6498	23	M	A	A	A	A	-	-	-	-	-	CA	-	SCA-3
67	VELUSAMY	6261	59	M	A	A	A	A	-	-	-	-	-	CI	CI	CS
68	VISHNU	49056	26	M	A	A	A	A	N	-	-	-	-	CTIS	-	CTIS
69	YUVARAJ	38651	25	M	P	A	A	A	-	-	-	-	-	CA	-	SCA -2
70	ZAMILA	58206	54	F	A	A	A	A	-	-	-	-	-	-	-	LAS

1. A : ABSENT
2. AC : ASTROCYTOMA
3. ACD : ALCOHOLIC CEREBELLAR DEGENERATION
4. ACM : ARNOLD CHIARI MALFORMATION
5. AL : ALCOHOL
6. AN : ACOUSTIC NEUROMA

7. AT : ATAXIA TELENGIECTASIA

8. AVM : ARTERIOVENOUS MALFORMATION

9. BM : BACETRIAL MENINGITIS

10. BSD : BRAIN STEM DEMYELINATION

11. BSH : BRAINSTEM HAEMORRHAGE

12. BSI : BRANSTEM INFARCT

13. BSS : BRAINSTEM STROKE

14. BST : BRAINSTEM TUMOUR

15. BTI : B/L THALAMIC INFARCT

16. C : CARBA MAZEPINE

17. CAT : CARBAMAZEPINE TOXICITY

18. CA : CEREBELLAR ATROPHY

19. CC : CORD COMPRESSION (SPINAL)

20. CD : CEREBELLAR DEGENERATION

21. CH	:	CEREBELLAR HAEMORRHAGE
22. CI	:	CEREBELLAR INFARCT
23. CVJ	:	CRANIOVERTEBRAL JUNCTION ANOMALY
24. CMS	:	COMPLETE MEDULLARY SYNDROME
25. CPAT	:	CP ANGLE TUMOUR
26. CS	:	CEREBELLAR STROKE
27. CSC	:	CERVICAL SPONDYLOTIC CHANGES
28. CTIS	:	CEREBELLITIS- TYPHOID
29. CT	:	CEREBELLAR TUMOUR
30. DM	:	DEMYELINATION
31. DRPLA	:	DENTATO RUBRO PALLIDO LUSIAN ATROPHY
32. E	:	EPTOIN
33. ET	:	EPTOIN TOXICITY

34. FA	:	FREDRIECH'S ATAXIA
35. GBS	:	GUILLAIN BARRE SYNDROME
36. HB	:	HAEMANGIO BLASTOMA
37. HSMN	:	HEREDITARY SENSORY MOTOR NEURONOPATHY
38. LAS	:	LABYRINTHINE ATAXIC SYNDROME
39. LMS	:	LATERAL MEDULLARY SYNDROME
40. MM	:	MEDIASTINAL MASS
41. N	:	NORMAL
42. NCS	:	NERVE CONDUCTION STUDY
43. P	:	PRESENT

44. PG	:	PONTINE GLIOMA
45. PN	:	PERIPHERAL NEUROPATHY
46. PNET	:	PRIMITIVE ECTODERMAL TUMOUR
47. PNS	:	PARANEOPLASTIC SYNDROME
48. SCA	:	SPINO CEREBELLAR ATAXIA
49. TFV	:	TRAUMATIC # OF VERTEBRA

S. No	Name	IP No.	Age	Sex	Family H/O	Drug H/O			CSF Analysis	Myelogram	NCS	X-Ray		CT SCAN	MRI	Diagnosis
						Al	E	C				Chest	Skull & Spinal			
21	EZILAN	59465	29	M	A	A	A	A	-	-	-	-	-	CA	-	SCA – I
22	FATHIMA	8642	36	F	A	A	A	A	DM	-	-	-	N	-	-	GBS
23	GAYATHRI	5616	49	F	A	A	A	A	-	-	-	-	-	BSI	-	LMS
24	HEMA	2264	17	F	A	A	A	A	-	-	-	-	-	N	-	CVJ
25	HEMANATHAN	89465	25	M	A	A	A	A	-	-	-	-	-	CA	-	FA
26	IYAPPAN	36086	24	M	A	A	A	A	-	-	-	-	-	CA	-	SCA -4
27	JAMILABANU	46612	54	F	A	A	A	A	-	-	-	-	-	BTI	-	BTI
28	JASMINE	94268	38	F	P	A	A	A	-	-	PN	-	-	-	-	HSMN
29	JOHNSON	4283	47	M	A	P	A	A	-	-	-		-	BSI	-	BSS
30	KANNAMAL	6086	40	F	A	A	A	A	-	-	-	MM	N	CD	-	PNS
31	KARUPUSAMY	4122	52	M	A	P	A	A	-	-	-	-	-	CD	-	ACD
32	KRISHNASAMY	2695	48	M	A	P	A	A	-	-	-	-	-	PNET	-	CT
33	KUMAR	50896	29	M	P	A	A	A	-	-	-	-	-	-	-	SCA -2
34	LATHA	52210	48	F	A	A	A	A	-	-	-	-	-	-	AN	CPAT
35	MANIAMAL	94680	40	F	A	A	A	A	-	-	-	-	-	AC	-	CT
36	MARIMUTHU	93422	33	M	A	A	A	A	-	-	-	+	-	CA	-	SCA -1
37	MYLAPPAN	85638	44	M	A	A	A	A	DM	-	-	-	-	-	-	CIDP
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62	THILAGAVATHI	8221	6	F	A	A	A	A	-	-	-	-	-	PG	PG	BST
63	VADIVU	962	49	F	A	A	A	A	-	-	-	-	-	BSI	-	BSS
64	VEERAKUMAR	9048	28	M	A	A	A	A	-	-	-	-	-	-	-	SCA-1
65	VELLAPPAN	2606	29	M	A	A	A	A	N	-	-	-	-	BSD	-	BSD
66	VELLINGRI	6498	23	M	A	A	A	A	-	-	-	-	-	CA	-	SCA-3
67	VELUSAMY	6261	59	M	A	A	A	A	-	-	-	-	-	CI	CI	CS
68	VISHNU	49056	26	M	A	A	A	A	N	-	-	-	-	CTIS	-	CTIS
69	YUVARAJ	38651	25	M	P	A	A	A	-	-	-	-	-	CA	-	SCA -2
70	ZAMILA	58206	54	F	A	A	A	A	-	-	-	-	-	-	-	LAS

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40. MM	:	MEDIASTINAL MASS
41. N	:	NORMAL
42. NCS	:	NERVE CONDUCTION STUDY
43. P	:	PRESENT

44. PG	:	PONTINE GLIOMA
45. PN	:	PERIPHERAL NEUROPATHY
46. PNET	:	PRIMITIVE ECTODERMAL TUMOUR
47. PNS	:	PARANEOPLASTIC SYNDROME
48. SCA	:	SPINO CEREBELLAR ATAXIA
49. TFV	:	TRAUMATIC # OF VERTEBRA